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FAMILIAL ADENOMATOUS POLYPOSIS

A PATIENT PERSPECTIVE ON LIFE AFTER SURGERY

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"the work that will do is that which is done by skilled hands,
guided by a clear mind and inspired by a loving heart"

inspired by Florence Nightingale

ABSTRACT

Familial adenomatous polyposis (FAP) is a rare hereditary condition with a high risk of gastrointestinal cancer. Individuals with FAP are committed to endoscopic screening surveillance throughout life in addition to having to undergo removal of the colon. With the intention to find strategies to develop care and support, the overall aim was to gain a deeper understanding of the life of surgically treated individuals with FAP. The first study used focus group interviews to collect data. The sample consisted of 14 surgically treated individuals, aged 18-75. Studies II-IV employed a cross-sectional design and were based on self-reported standardized questionnaires to measure abdominal symptoms, health-related quality of life, illness perceptions and quality of care. Assessments were performed using the Abdominal Symptom Questionnaire (ASQ), the Medical Outcomes Study Short Form 36 Health Survey (SF-36), the Revised Illness Perception Questionnaire (IPQ-R) and the Quality of Care from the Patients Perspective (QPP). The sample consisted of 209 surgically treated individuals, aged 18-75 (response rate 76%). The focus group interviews revealed that surgically treated individuals' have several concerns, including issues about hereditary, the life-long nature of their illness and abdominal symptoms. Individuals use different strategies to minimize the effects of their illness on everyday life. The results from the self-reported questionnaires revealed that 91% of the individuals perceived at least one abdominal symptom during the past three months and reported a mean of 7 (SD 4.61; range 1-18) symptoms. A high number of abdominal symptoms predicted poorer health status. Furthermore, individuals perceived FAP as chronic and to a low extent possible to control. Negative consequences of the illness predicted a worse HRQL as did number of abdominal symptoms. Individuals receiving specialized care and those with a low number of abdominal symptoms were more likely to perceive the quality of care they received as better. In conclusion, to improve HRQL abdominal symptoms and negative consequences on life should be monitored and self-care supported when caring for surgically treated individuals with FAP. Care should be provided by a team of health-care professionals specialized in FAP and is recommended to be coordinated by a nurse. Such care should be holistic and focus on supporting the self-care of the individual.

Keywords: Abdominal symptoms; Chronic illness experience; Colorectal surgery; Familial adenomatous polyposis; Heredity; Health-related quality of life; Illness perception; Quality of care

LIST OF PUBLICATIONS

The thesis is based on the following papers which are referred to by their Roman numerals:

I. Fritzell K, Persson C, Björk J, Hultcrantz R, Wettergren L.
Patients' Views of Surgery and Surveillance for Familial Adenomatous Polyposis. Cancer Nursing. 2010; 33(2)E17-23.

II. Fritzell K, Eriksson LE, Björk J, Hultcrantz R, Wettergren L.
Self-Reported Abdominal Symptoms in Relation to Health Status in Adult Patients With Familial Adenomatous Polyposis.
Diseases of the Colon and Rectum. 2011;54(7)

III. Fritzell K, Eriksson LE, Björk J, Sprangers M, Wettergren L.
Illness Perception in patients with hereditary colorectal cancer.
Submitted.

IV. Fritzell K, Eriksson LE, Björk J, Sprangers M, Wettergren L.
Patients with genetic cancer undergoing surveillance at a specialized clinic rate quality of care better than patients undergoing surveillance at other clinics. In manuscript.

CONTENTS

LIST OF ABBREVIATIONS

ANOVA	Analysis of variance
APC	Adenomatous polyposis coli (gene)
CSM	Common sense model of self-regulation
DNA	Deoxyribonucleic acid
EGD	Esophagogastroduodenoscopy
FAP	Familial adenomatous polyposis
GERS	Gastroesophageal reflux symptoms
HRQL	Health-related quality of life
IBS	Irritable bowel syndrome
IPAA	Ileal pouch anal anastomosis
IRA	Ileorectal anastomosis
MAP	MUTYH-associated polyposis
MCS	Mental component summary
PCS	Physical component summary
MUTYH	MutY Homolog (gene)
ORs	Odds ratio
SD	Standard deviation

1 INTRODUCTION

Familial adenomatous polyposis (FAP) is a hereditary illness that, without prophylactic removal of the colon, is associated with an almost 100% risk of developing cancer. Individuals with FAP are free of symptoms before surgery, but often perceive symptoms related to impaired bowel function after surgery. Prophylactic surgery is therefore commonly expressed as the time point when individuals with FAP realize that they actually have an illness. When I was employed at the national Swedish Polyposis Registry (a competence center for hereditary polyposis syndromes, including a registry database and an outpatient clinic), in 1999, I had provided care for a few individuals with FAP at the surgery ward where I previously worked. Since I started to work at the Swedish Polyposis Registry I have provided care for approximately 100 surgically treated individuals with FAP. In addition, I have recorded all data into the Swedish Polyposis Registry, which means that I have read thousands of medical records the last ten years for additionally 250 individuals. Finally, as the coordinator of the Swedish Polyposis Registry, I have been communicating with persons (patients, relatives and health-care providers) throughout the country. All this has given me an insight into how it can be to live with FAP, as well as uninvestigated areas that seemed to affect an individuals' life. Issues of concern were related to care given by health care providers who have no or little knowledge of FAP, the lifelong endoscopic surveillance program and non-specific abdominal symptoms not previously investigated. I found that care for surgically treated individuals with FAP was not similar for all persons, i.e. it seemed that care was more dependent on where in the country the person lived than on the actual care needed. At the planning stage of this thesis, there was consensus in the Western world regarding the medical management of FAP. However, consensus regarding the management of an individual's everyday life after surgery, including symptom management, was not available because of a lack of research in this area. Furthermore, there were no Swedish reports published on psychosocial concerns in surgically treated individuals with FAP and there were almost no international publications. Nevertheless, it was generally accepted that individuals with FAP suffer from symptoms related to bowel dysfunction after surgery though knowledge as to how the symptoms affect life was limited. During the work of this thesis, studies that have been published are mainly in the field of genetic counseling and about young adults with FAP. In addition, one thesis studied the psychosocial and behavioral impact on FAP.

2 CHRONIC ILLNESS EXPERIENCE

Defining chronic illness is complex in that it is dependent on the person's experience of an illness. Concepts included in the chronic illness experience vary. For instance, researchers with a psychology background will base their research on psychological implications associated with the illness, whereas sociologists will focus on behavioral implications while anthropologists investigate the experience of chronic illness in a social, cultural and ethnic context (1). Many of the criteria used to define chronic illness experience overlap each other and the differences are more a matter of the language used. Further, the chronic illness experience is too complex to define easily because of the different ways that individuals identify their chronic illness. Just because two individuals have the same chronic disease, does not necessarily mean they view the illness similarly (e.g., one of them may not even view his or her disease as chronic) (1). One definition that takes the illness experience into account from a nursing perspective was formulated by Curtin and Lubkin (2), who stated, *"Chronic illness is the irreversible presence, accumulation, or latency of disease states or impairments that involve the total human environment for supportive care and self-care, maintenance of function, and prevention of further disability".* In the context of nursing, chronic illness experience is further based on a holistic and patient-centered view, which is based on the understanding of patients' illness experiences (3). Criteria usually included in the chronic illness experience are lack of control, self-care (4), adaptation, powerlessness, social isolation, sexuality, quality of life and adherence (5). In addition, how individuals view themselves as chronically ill is dependent on the family and the social and health care context (1, 5). Self-care has been described as taking control over the disease, with the aim of living a life as normal as possible. To stay in control individuals monitor their body for signs of illness, or what Thorne and colleagues have called, "body listening" (4). Although there is no uniform definition of self-care, Hertz (6) developed a working definition based on diverse perspectives of the concept. The working definition states, "self-care is self-determined, unique to each individual based on that person's life experience, values, beliefs, and personal characteristics and abilities, including biopsychosocial-spiritual and functional abilities. Self-care includes a variety of activities, including adherence to prescribed medical treatments and guidelines for disease prevention, as well as pursuing personal interests that promote well-being (6).

2.1 SYMPTOM EXPERIENCE

The word symptom can be traced to its Latin origin, "*synthoma"* meaning 'sign of something evil' (7). Today, symptom is defined as "a change in your body or mind that shows that you are unhealthy" (8) and further, a private experience unknown by others (9). An individual's experience of a symptom is thought to include symptom occurrence, frequency, intensity, distress and meaning of symptoms, including the situational and existential meaning of individual symptoms as well as the impact of symptom clusters (10). A symptom experience is influenced by antecedents, such as demographic (e.g., age, gender and marital status) and individual characteristics (e.g., health knowledge, values and experiences) and disease characteristics (e.g., type of treatment and clinical factors) (10). In the context of FAP symptoms (which are caused by the prophylactic colorectal surgery aiming to prevent colorectal cancer) that are related to an impaired bowel function are an everyday concern (11).

2.2 ILLNESS REPRESENTATION

Individuals with a chronic illness are considered to adapt an illness behavior that is needed to improve or maintain health, as well as to accommodate their own and societal expectations. In the common sense model of self-regulation (CSM) constructed from in-depth interviews with patients to elicit illness representation, symptoms are the individual's concrete representations of illness (12). Three principles underlie CSM: 1) individuals are problem solvers who seek information and adequate actions, 2) the individual's illness representation guides actions (coping) and evaluation of action outcomes and 3) illness perceptions are individual and may not necessarily correspond to medical facts (13). When persons perceive a health threat (e.g., a media message about swine flu or symptoms of an illness), they have to deal with two phenomena: the individual's perception of a health threat and the emotional reactions to the threat. The two phenomena operate in parallel and are partly independent in that they may motivate the same behavior or may motivate contrasting behaviors (13, 14). The illness representation evolves and will be more detailed and rich with increasing experience and knowledge, which results in a more reliable influence on the patient's behavior (15). The Revised version of the Illness Perception Questionnaire (IPQ-R), which has been used in patients with a variety of diagnoses, was developed to assess illness perception (16).

2.3 QUALITY OF CARE

Health care is integrated with the individuals' self-care actions because quality of care is a part of their self-care recourses (6). Good outcomes are believed to be a result of productive interactions between the individual and the health care providers, where both the individual and the health care providers need to be active (17, 18). It has been suggested that nurse practitioners or specialized nurses are able to assume a leadership role in the health system transformation for chronic disease care because nursing's disciplinary is based on a holistic, individualized care (6, 18). A model that considers the patient-provider relationship is in line with the framework of Wilde and colleagues (19) in which quality of care from the patients' perspective is underlined. Based on interviews with adult hospitalized patients, they formulated a model in which quality of care could be understood. The model comprises *the patient's preferences and qualities related to the physical and administrative care environment*. The individuals' preferences refer to rational aspects (e.g., their ambition for predictability and calculability in life) and human aspects (e.g., their expectations that their unique situation will be taken into account). The qualities related to the physical and administrative care environment refers to person-related qualities of the caregivers and infrastructural components (e.g., organizational rules and technical equipment) (19). Individuals' perception of quality of care can be considered from four dimensions: medical-technical competence, physical-technical conditions, identity-oriented approach and socio-cultural atmosphere. Medical technical competence refers to the patient's wishes that caregivers possess a high level of medical-technical competence, i.e. qualified personal who can make correct diagnoses, propose relevant examinations and alleviate symptoms. It also includes wishes of having access to caregivers with a high level of medical-technical competence. Physical-technical conditions refer to the availability of a care organization providing the necessary physical-technical conditions, such as a clean, comfortable and safe environment, as well as advanced medical-technical equipment. The identity-oriented approach denotes the human aspect of the patient's relationship to caregivers, such as the wish of qualified caregivers with an interest in the patient as a unique person as well as a wish for mutual understanding, respect and collaboration between caregivers and patient. The socio-cultural atmosphere pertains to a human environment that includes a care organization that allows an individual's wishes and needs to be prioritized before fixed routines and where the individual can choose to socialize or not depending on mood and state of

mind (19). The model has been operationalized to be assessed with the questionnaire Quality of Care from the Patients' Perspective (20).

2.4 HEALTH-RELATED QUALITY OF LIFE

In 1948, the World Health Organization (WHO) (1948) presented a broad definition of health: "a state of complete physical, mental and social well-being and not merely the absence of disease or infirmity". A definition provided in the context of self-care in chronic illnesses is in line with the WHOs definition (21). Hertz (6) stated that "health is holistic, meaning that it is more than the absence of physical and mental health" and that "health and chronic illnesses can coexist". Health-related quality of life (HRQL) is an outcome measure to evaluate the impact of an illness on an individual, assess response to treatment and guide interventions (6, 22). Although no consistent definition exists, general health, physical functioning, physical symptoms, emotional functioning, cognitive functioning, role function, social well-being and sexual functioning are areas generally included in HRQL (23). Eight health concepts provided by Ware and colleagues (24) will be referred to as HRQL in this thesis: 1) limitations in physical activities because of health problems, 2) limitations in social activities because of physical or emotional problems, 3) limitations in usual role activities because of physical health problems, 4) bodily pain, 5) general mental health (psychological distress and well-being), 6) limitations in usual role activities because of emotional problems, 7) vitality (energy and fatigue) and 8) general health perception. Given the broad concept of health and HRQL the measurement of the concepts varies (23). The most widely used questionnaire is the Short-Form Health Survey (SF-36), which was designed to assess generic health concepts that are not specific to age or disease (24).

3 BACKGROUND

3.1 HISTORICAL OUTLINE

In 1953, at St Erik's Hospital in Stockholm 'diffuse adenomatosis' was suspected in a 44-year-old male who underwent surgery for colon cancer. The individual had a desmoid tumor (locally infiltrating and occasionally life-threatening lesion) and epidermoid cysts known to be associated to clinical manifestations with, what later would be named, 'familial adenomatous polyposis'. The individual was finally given the correct diagnosis in 1961(25). Inspired by the work of Dukes (26), the first attempts were made to trace Swedish families with FAP. Finally, in 1959, the Swedish Polyposis Registry at St Erik's Hospital was established (25). The primary aim of the registry was to prevent colorectal cancer in persons with FAP in the family (26). The organization and outline of the registry were based on the work of the Polyposis Registry at St Mark's Hospital in London, which was set up already in 1925 (27). The familial occurrence of the disease was observed already in the latter half of the 19th century, but the heritage was first verified in 1952 by Dukes (26) when he analyzed a larger number of families. Two theses have been conducted at the Swedish Polyposis Registry. The first thesis was undertaken by the physician Thor Alm (25) with the aim of gathering all individuals with FAP in Sweden to calculate incidence, set up a registry and evaluate the results of surgical treatment. At that time, the Swedish Polyposis Registry included 80 fully investigated families (25). The second thesis, by the physician Jan Björk, included the evaluation of the efficiency of the national screening program in reducing the risk of colorectal cancer in individuals with FAP. The results are based on data between 1957 and 1996 (28). In 1996, the number of families with FAP in the registry had increased to 145 (29). The present thesis contributes to the knowledge of FAP by increasing the understanding of the impact on the lives of surgically treated individual's with FAP.

3.2 GENETICS

DNA (Deoxyribonucleic acid) contains all information needed when building a person. A mutation, which is the cause of genetic diseases, is a change in the individual's DNA. If the DNA is the sentence that gives information about how to build, the mutation can be seen as a spelling error. A spelling error (mutation) can give the sentence a different meaning, which may cause illness. FAP is inherited in an autosomal dominant way,

which means that a child of a parent with FAP has a 50% risk of inheriting the syndrome (29) (Figure 1). The gene was identified in 1991 and is called the adenomatous polyposis coli (APC) gene (30). In 2000, another polyposis gene, the MUTYH gene was identified. This form of polyposis is called MUTYH-associated polyposis (MAP) and the mode of inheritance is autosomal recessive (31). This means that for a child with a parent with MAP the risk of inheriting the syndrome is approximately 0.25% since the frequency of heterozygous mutation carriers in the background population is about 1/100. (31). Molecular genetic testing has failed to find disease-causing mutation in approximately 10% of the Swedish polyposis families with clinical evidence of disease, indicating that more genes might be involved in causing the illness. In addition, approximately 10% of all FAP individuals have been reported to be “de novo” without clinical or genetic evidence of FAP in the parents (29).

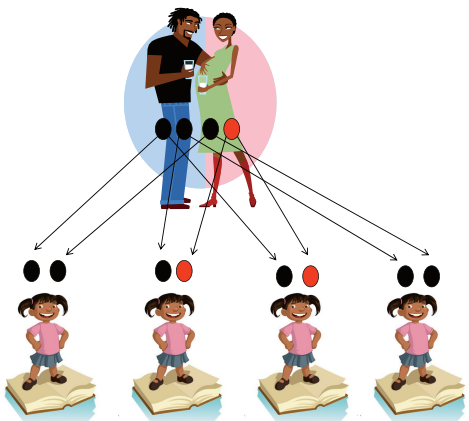

Figure 1. Autosomal dominant inheritance. When having a parent with FAP (regardless of gender) the child has 50% risk of inheriting the disease.

3.3 INCIDENCE AND PREVALENCE

FAP is responsible for approximately 0.1% of all colorectal cancer cases. In Sweden, about 350 persons live with FAP, yielding a prevalence of 35 per 1000 000 inhabitants. Annually, 7-8 children are born with FAP in Sweden (29). The incidence and prevalence of FAP are similar in the Western world (32). Because our knowledge of MAP is relatively new, incidence and prevalence have not been fully investigated.

3.4 CLINICAL CHARACTERISTICS

FAP is characterized by the development of hundreds to thousands of adenomas in the colorectum (33). Mortality from colorectal cancer is assumed to be almost 100%

without treatment (colorectal surgery), and half of the individuals diagnosed with FAP would die of cancer before the age of 50 (34). There is also an increased risk of cancer in the duodenum (35). An increased risk of hepatoblastoma, as well as thyroid cancer and medulloblastom in individuals with FAP has also been reported (32). Furthermore, FAP is associated with extraintestinal manifestations, such as desmoid tumours (locally infiltrating and occasionally life-threatening lesions mostly located in the abdominal cavity or abdominal wall), epidermoid cysts (benign cysts appearing as a bump on the skin), cutaneous lipomas and osteomas (32). The characterization of MAP is similar to FAP, although MAP is considered less severe than FAP in age of onset, numbers of colorectal adenomas and extraintestinal manifestations (32, 36). Because our current knowledge about the nature of MAP is limited, treatment and follow-up of individuals with MAP are similar to FAP in Sweden (36). Henceforth, in this thesis all individuals will be referred to as patients with FAP.

3.5 DIAGNOSIS

All individual with a risk of FAP, with the exception of families in which no mutation is found, are offered genetic testing (blood sample) to confirm or deny a diagnosis of FAP (32). For children with a history of FAP in the family, screening starts at the age of 10-12 years with a genetic test or with colonoscopy if a genetic test is not possible (32). Children with a verified diagnosis, either because of the finding of a disease causing mutation or by identified adenomas in the colon or rectum, will be followed annually with colonoscopy until prophylactic colorectal surgery is performed (32).

3.6 GENETIC COUNSELING

All genetic testing shall be preceded by genetic counseling, which is delivered by different health professionals (e.g., physicians, nurses, psychologists) educated in genetic counseling. In Sweden, most genetic counselors are affiliated to a genetic clinic at University Hospitals. A genetic counseling session includes the following: taking family medical history, providing information about the inheritance and risk for family members, collecting blood samples for testing and processing result disclosure. It is important that genetic counselors support the individual to make an informed choice about testing, so that individuals know what a positive or negative result means for them and for close family members (37). Testing for genetic illnesses is associated with concerns and can be stressful before test disclosure (37). However, there are both positive and negative aspects of genetic testing when interviewing young people who

had undergone genetic testing for FAP (38). Participants felt empowered and family relationships were improved (38); however, lack of control and involvement in the decision to be tested were also discussed (38, 39). Another important function for genetic counselors is to support individuals in family planning. Having FAP has been found to influence an individual's desire to have children. However, termination of pregnancy has not been seen as an alternative (40, 41), whereas a person is more positive to prenatal diagnosis (test for disease in a embryo before it is born) in order to prevent having a child with FAP (41). Furthermore, parents have been found to prefer FAP testing in children at birth or early childhood instead of waiting 10-12 years according to guidelines (40, 41). Genetic counseling is recommended at time for genetic testing (32), although regular follow-up in young adults after test disclosure has been suggested in order to provide support, especially in family planning (40).

3.7 TREATMENT AND OUTCOMES

3.7.1 Prophylactic surgery

To prevent colorectal cancer prophylactic surgery is normally performed between the ages of 18 and 20 years, with removal of the colon (colectomy) or colorectum (proctocolectomy). The two most common prophylactic colorectal surgical procedures in individuals with FAP are ileorectal anastomosis (IRA) and ileal pouch anal anastomosis (IPAA). When IRA is performed, the large intestine is removed and the ileum is anastomosed to the rectum. To create an IPAA, both the large intestine and rectum are first removed and the last part of the ileum is turned into a pouch, which is sewn onto the anus (42). A third, less common, method of surgery to prevent colorectal cancer in FAP individuals is through the creation of a permanent ileostomy. In ileostomy the large intestine is removed and the ileum is then attached to the abdominal wall (stoma) (43). The lifetime risk of additional operations (IPAA or permanent ileostomy) because of advanced polyposis or cancer in the remaining rectum or pouch is high, up to approximately 70% (30). There is also a risk that individuals will need further surgery because of polyposis or cancer in the duodenum (34). Preliminary data from the Swedish Polyposis Registry show that by the age of 60, 15% of the individuals have had their duodenum prophylactic removed.

3.7.2 Endoscopic surveillance

Prophylactic surgery is followed by lifelong endoscopic surveillance. Individuals with IRA undergo examination of the rectum every 6 months, whereas patients with IPAA undergo examination of the pouch every 24 months (32). The procedures are similar: a long, flexible, lighted tube is inserted into the intestine through the anus and the lining of the intestine is visible in a TV monitor to all persons in the room. Removal of polyps is possible since the endoscope permits passage of an instrument with forceps at the end. To perform a high quality examination the intestines need to be empty before the examination. Thus, individuals need to prepare with bowel cleaning prior to the examination. Because of the risk of severe polyposis or cancer in the duodenum, investigation of the stomach and the duodenum (esophagogastroduodenoscopy, EGD) is recommended from the age of 25 (32, 35). EGD is a procedure in which the patient swallows a thin, flexible, lighted tube, which allows viewing the lining of the esophagus, stomach and duodenum on a TV monitor. To perform a safe and high quality examination at the same time the stomach should be empty. Consequently, the individual needs to be without food and drink six hours before the procedures. Surgically treated individuals with FAP are probably those that undergo most endoscopic examinations during their life (e.g., an individual with IRA undergoes examination of the rectum every 6 months). In addition, examination of the stomach and duodenum (EGD) is performed every 24 months or even more frequently depending on the examination findings. Endoscopic examinations in patients are often associated with discomfort and anxiety. However, the impact of frequent endoscopic surveillance, as in the case with surgically treated individuals with FAP, has not been investigated. Because of the high risk of cancer, it is important that individuals with FAP consider recommended endoscopic intervals (44). Yet, knowledge about adherence among surgically treated individuals with FAP is limited. One study found that non-adherence to endoscopic screening in individuals with an IRA was 26% (45). Another study found that 48% of individuals with a positive mutation for FAP or individuals who had undergone prophylactic surgery to be non-adherent (46). Suggested factors related to non-adherence are individuals' perception of having a low risk of cancer, lack of provider recommendations, deficiency or absence of health insurance (46), fear of additional surgery, social inconvenience and young adulthood (44).

3.7.3 Complications related to surgery

Individuals with FAP are normally asymptomatic before surgery, but it is well known that bowel function is impaired after surgery (11). Reported symptoms related to impaired bowel function are increased defecation frequency, nighttime defecation, leakage, difficulty in distinguishing gas from feces, perianal skin problems, symptoms of small bowel obstruction and dietary restrictions (11, 47, 48). Individuals with their rectum removed (IPAA) often report more symptoms than those who have undergone colectomy without removal of the rectum (IRA) (11, 49). Further, knowledge of perceived bowel function in individuals with an ileostomy is limited (50). Nevertheless, one study showed that individuals with an ileostomy perceived having an excellent bowel function; however, the result was based on interviews with only 10 individual (50), which must be considered as a limitation. In addition, knowledge of an individual's perception of bowel function, as well as what abdominal symptoms trouble them after removal of the duodenum is limited, although pain has been reported (51). Furthermore, impairment in sexual function, such as diminished libido and/or erection in males and dyspareunia in females, has been reported (48). Moreover, fecundity in women has been found to decrease after IPAA surgery, whereas no impact on fecundity in women with IRA has been reported (52).

3.7.4 Health-related quality of life in individuals with FAP

Quality of life research in individuals with FAP has mainly focused on the impact of bowel function and surgery. Efforts have been made to determine which type of surgery is most beneficial for the individual. However, inconsistent results have been found, making it difficult to recommend one type of surgery over another (11). Quality of life in surgically treated individuals with FAP has been reported to be in parity with population norms (47-49, 53,54). Nevertheless, some studies have found associations between quality of life (53), or aspects of quality of life (34, 55), and bowel function. Further, different effects on specific aspects of quality of life have been noted, such as physical functioning, changes in body image, employment and sexual impact (54-56). One study indicates that individuals with FAP and desmoid tumor perceive a reduced quality of life (57). Moreover, surgical complications and comorbidity have been found to be associated with reduced quality of life (54). Further, guilt regarding the risk of passing on the disease to their offspring (58), fear and anxiety about future health because of the risk of cancer has been reported (58, 59). FAP related stress has been

found in a minor group of individuals (55, 59), why a screening questionnaire to identify individuals who need support has been recommended (59). In addition, supportive counselling is considered important both before and after surgery (54, 55).

3.8 FAP CARE IN SWEDEN

The organization of the Swedish Polyposis Registry consists of a computerized database, the outpatient clinic and the endoscopy unit. Today, the Swedish Polyposis Registry includes 240 families (236 with FAP and 4 with MAP). Of the 600 persons in the registry with FAP (or MAP), 350 were still alive at the time of the present study. Data on family history, genetic status, extraintestinal manifestations, surgery and endoscopic examinations, including pathology reports, are included in the registry. Registry data are obtained from the patient medical records. The Swedish Polyposis Registry is a database for research, and assists with family investigations and information to patients, relatives and health care providers throughout the country. In addition, the registry organizes a yearly nationwide education day for patients and relatives. At the time of the present studies, the outpatient clinic that is affiliated to the Swedish Polyposis Registry provided specialized care based on a team of one consultant and one specialized nurse; the latter also serves as a genetic counselor. Since then, the team has expanded to include two consultants and two nurses. One important aim is to provide continuity and individualized care by coordinating and performing all endoscopic examinations and providing individualized regular appointments to consultants, nurses and the genetic counselor. The outpatient clinic coordinates care for approximately 30% (n=110) of the Swedish population with FAP. The primary catchment area is Greater Stockholm with a population of approximately 2 million inhabitants; health care for most of the remaining FAP individuals in Sweden is provided by the hospital in the county council where the patient resides. At time of this study, individuals included in the study (n=153) from outside the Swedish Polyposis Registry clinic were managed by approximately 75 physicians (median=1, range 1-6) at 34 hospitals (7 university hospitals; data obtained from the Swedish Polyposis Registry). When scrutinizing medical records for those individuals, organization of the care process was found to vary between hospitals. The outpatient clinic at the Swedish Polyposis Registry offers second opinion consultations and genetic counseling for individuals outside its catchment area. A referral from their county council is needed,

however. The possibility for an individual with FAP to have a referral for a second opinion varies.

4 THESIS RATIONALE

Previous studies investigating the influence on life in surgically treated individuals with FAP have focused on symptoms related to impaired bowel function. The studies have been limited to two of three surgical procedures that are usually performed to prevent colorectal cancer. The sample sizes are often small and typically include heterogeneous samples (e.g. patients FAP and inflammatory bowel disease). Furthermore, findings from studies pertaining to quality of life and psychosocial issues are often based on samples with a mix of surgically treated individuals with FAP, patients diagnosed with FAP without surgery and individuals with a risk of FAP. Moreover, knowledge of how surgically treated individuals perceive their illness is therefore limited (e.g., HRQL, abdominal symptoms, endoscopic surveillance) and no studies have been undertaken to investigate how individuals perceive the quality of care.

5 AIM

With the intention to find strategies to develop care and support, the overall aim of this thesis was to gain a deeper understanding of the lives of surgically treated individuals with FAP. This overall aim was approached with the following specific aims:

I. To gain a deeper understanding of how FAP affects life by exploring patients' views of what it is like living with the condition and having to be committed to a lifelong screening program.

II. To investigate self-reporting by adults with FAP with regard to presence, frequency and troublesomeness of abdominal symptoms in relation to health status. An additional aim was to study abdominal symptoms in relation to gender and type of colorectal surgery.

III. To examine how patients with FAP perceive their illness and experience HRQL. Additional aims were to examine how illness perceptions and HRQL may differ between subgroups of patients in gender, family history of FAP and adherence to treatment. A further aim was to investigate whether any socio-demographic and clinical variables (including number of abdominal symptoms) and illness perceptions are associated with HRQL.

IV. To test the hypothesis that surgically treated patients with FAP who receive specialized care rate quality of care more favorably than patients who receive non-specialized care. An additional aim was to investigate whether any socio-demographic and clinical variables are associated with quality of care.

6 METHODS

6.1 DESIGN

Study I is a descriptive qualitative study based on data obtained by focus group interviews. Studies II-IV used a cross-sectional, cohort design. Study outcomes, participants, data collection and analyses are presented in Table 1.

Table 1. Overview of outcomes, participants, data collection and analyses within this thesis

Study	Outcomes	Participants	Data collection	Analyses
I	Patients view of what it is like living with FAP	14 patients, age 28-67	Focus group interviews	χ^2 tests, Descriptive content analysis
II	Number, frequencies and troublesomeness of abdominal symptoms Abdominal symptoms related to bowel function Health Status Factors related to Health Status	209 patients, age 18-75; 93 men and 116 women	The Abdominal Symptom Questionnaire (ASQ) Five additional abdominal symptoms from the Revised Illness Perception Questionnaire (IPQ-R) The Short Form 36 Health Survey (SF-36)	χ^2 tests, Independent t-tests, One-way ANOVA, Pearson correlation coefficients, 3-step hierarchical regression analyses
III	Illness perception Health Related quality of life (HRQL) Factors associated with HRQL		The Revised Illness Perception Questionnaire (IPQ-R) The Short Form 36 Health Survey (SF-36) The Abdominal Symptom Questionnaire (ASQ)	χ^2 tests, Independent t-tests, Cronbach's alpha coefficients, 3-step hierarchical regression analyses
IV	Quality of Care Factors related to Quality of Care		The Quality of Care from the Patients Perspective (QPP) The Abdominal Symptom Questionnaire (ASQ)	χ^2 tests, logistic regression analyses

6.2 PARTICIPANTS

6.2.1 Study I

Participants were recruited from the outpatient clinic at the national Swedish Polyposis Registry, Karolinska University Hospital, Stockholm. Forty-three of 90 patients fulfilled the criteria for participation. Inclusion criteria were aged 18-75 years, having a mutation in the adenomatous polyposis coli gene (APC) or meeting the criteria for classic FAP. Fourteen of the 43 patients invited agreed to participate (Table 2).

Table 2. Participants' characteristics (n=14)

	mean (range)	n
Age	47 (28-67)	
Sex		
Women		10
Men		4
Marital status		
Married/partnered		10
Single		4
Participants having children		9
Socio-economic status		
Employed		7
Student		1
Disability pension*		2
Retired		4
APC mutation		14
New mutation		2
Median time from diagnosis to study	25 (4-47)	
Type of surgery		
IRA		6
IPAA		4
Permanent ileostomy		4
Median time from surgery to study	24 (4-44)	
Second surgery		
Permanent ileostomy		2
Upper gastrointestinal surgery		
Duodenectomy		2
Desmoid tumour		1
FAP associated cancer		3

* Not due to FAP

6.2.2 Studies II-IV

Participants were recruited from the national Swedish Polyposis Registry, Karolinska University Hospital, Stockholm. Patients eligible for inclusion were aged 18-75 years, having a mutation in the APC, or homozygote mutation in the MUTYH gene, or who met the criteria for classic FAP and who had prophylactic colorectal surgery. Two hundred seventy-six patients met eligibility criteria and thus were invited to participate in the study. One patient did not speak Swedish and was therefore excluded. A flow chart describing participants and non-participants is shown in Figure 1. Of 275 patients, 209 (76%) consented to participate (Table 3).

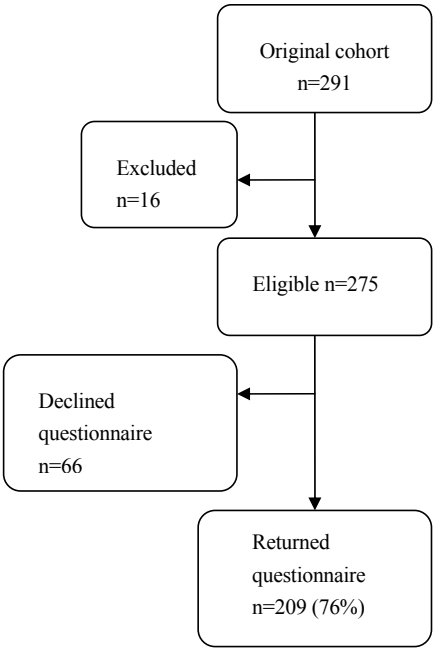

Figure 1. Flow chart describing participants and non-participants

Table 3. Participants' characteristics (n=209)

	mean (SD; range)	n	%
Age	49 (14; 18-75)		
Sex			
Men	48 (15; 18-75)	93	44
Women	49 (13; 23-74)	116	56
Living situation			
Partnered		139	67
Single		61	29
Living with parents or other		9	4
Occupational status			
Working		123	59
Student		12	8
Retired		42	20
Disability pension		17	8
Sick-listed		11	5
Participants having children		144	69
Educational attainment			
Compulsory school		56	27
High-school (2 years)		37	18
High-school (3 years)		33	16
High-school (4 years)		7	3
Post graduate school		27	13
University		45	22
Family history of FAP[a]		169	81
Mutation			
APC		158	76
MYH		7	3
Mutation not found		23	11
Not tested		21	10
Age at diagnosis	26 (12; 3-57)		
Age at first colorectal surgery, years	28 (11; 5-58)		
Time since first colorectal surgery to study, years	21 (12; 1-50)		
Times since last colorectal surgery to study, years	14 (10; 1-50)		
First colorectal surgery			
IRA[b]		126	60
IPAA[c]		47	23
Ileostomy		22	11
Continent ileostomy		10	5
Other		4	2

Table 3. continued	**n**	**%**
Second colorectal surgery		
IPAA	39	19
Ileostomy	22	11
Content ileostomy	6	3
Third colorectal surgery		
Ileostomy	4	2
Colorectal surgery at time for study		
IRA	71	34
IPAA	82	39
Ileostomy	39	19
Continent ileostomy	14	7
Other	3	1
Total number of colorectal procedures		
One	132	63
Two	66	32
Three or more	11	5
Upper gastrointestinal surgery	12	6
Whipple procedure	11	5
Duodenectomy		
Desmoid tumor	20	10
FAP related cancer		
Intestinal cancer	29	14
Extraintestinal cancer	11	5
Intestinal and extraintestinal cancer	10	5
Adherence to endoscopic surveillance		
Adherent	180	86
Non-adherent[d]	29	14
Organization of care		
Specialized care[e]	57	27
Non-specialized care	152	73

[a] Participants has family members with FAP
[b] IRA=ileorectal anastomisis
[c] IPAA=ileal pouch anal anastomosis
[d] No record of endoscopic surveillance one year later than recommended intervals
[e] The outpatient clinic affiliated to the national Swedish Polyposis Registry, Karolinska University Hospital

6.3 DATA COLLECTION

6.3.1 Focus group interviews (Study I)

In study I data were collected using focus group interviews. Issues described in the literature as affecting the lives of patients with FAP as well as topics recognized in practice as important were formed into possible statements and questions to guide discussions in the event patients themselves did not raise any issues (Figure 2). The identified areas were FAP's impact on the life of the patient, hereditary aspects and the effects of prophylactic colorectal surgery and endoscopic surveillance on FAP patients.

6.3.2 Instruments (Studies II-IV)

The Abdominal Symptoms Questionnaire (ASQ) was used to measure abdominal symptoms (60) (Studies II-IV). The original ASQ is divided into three parts, of which only the first part was used in this thesis. This part includes 21 items measuring the presence of general abdominal symptoms. Respondents were asked to indicate the presence or absence (yes/no) of selected symptoms over the past three months as well as their frequency ("every day", "every week" or "every month"). The respondents were also asked to rate the troublesomeness of the symptoms on a seven-point Likert-style scale with the anchors "mild" and "very severe". The complete ASQ was originally designed to identify three functional gastrointestinal disorders: dyspepsia, gastroesophageal reflux symptoms (GERS) and irritable bowel syndrome (IBS). The instrument has shown acceptable sensitivity and specificity in identifying persons with dyspepsia and IBS (60). In the present study troublesomeness of symptoms was calculated by adding the score (possible score 1-7) for all symptoms and dividing the sum by the number of reported symptoms. Example of question format and responses available in the ASQ (Figure 2)

The Abdominal Symptom Questionnaire (ASQ)
Have you been troubled by abdominal distension during the last three months?

Yes ☐ No ☐
If yes:
Every day ☐ Every week ☐ Every month ☐

Severity:
Mild ☐ ☐ ☐ ☐ ☐ ☐ ☐ Very severe

Figure 2. Example of question format and responses available in the ASQ

The SF-36 was used to measure HRQL (24). The instrument consists of 35 items measuring eight dimensions: Physical functioning (PF), Role-physical (RP), Bodily pain (BP), General health (GH), Vitality (VT), Social functioning (SF), Role-emotional (RE) and Mental health (MH). In addition, one single item determines perceived differences in health status over the past year. Response choices vary from two to six. Twenty items refer to the four previous weeks, one concerns health transition over the past year and fifteen pertain to the present situation. Raw scores for each item were coded, summed and transformed into a scale from 0 (worst possible health status) to 100 (best possible health status) following standard SF-36 scoring algorithms (24). Based on the eight scales, two summary scales have been constructed for physical and mental health: the physical component summary (PCS) and the mental component summary (MCS). The PCS is primarily a measure of PF, RP, BP and GH, whereas the MCS mainly encompasses VT, SF, RE and MH (61).The Swedish version has shown satisfactory results for reliability and validity (61, 62).

The Revised Illness Perception Questionnaire (IPQ-R) was used to measure the participants' beliefs and understanding of their illness perceptions (16). The instrument includes nine components: Timeline-chronic (6 items; e.g., "My condition is likely to be permanent rather than temporary"), Timeline-cyclic (4 items; e.g., "My symptoms come and go in cycles"), Consequences (6 items; e.g., "My condition has major consequences on my life"), Personal control (6 items; e.g., "The course of my condition depends on me"), Treatment control (5 items; e.g., "My treatment will be effective in curing my illness"), Emotional representation (6 items; e.g., "My illness makes me feel afraid "), Illness coherence (5 items; e.g., "My condition is a mystery to me"), Causes (18 items), which measures a patient's idea about the etiology of his or her condition (psychological, risk factor, immune and accident/chance), and an Identity component (see below). All items of the IPQ-R are rated on a five-point Likert scale ranging from "strongly agree" to "strongly disagree", except the Identity component. Higher scores on the Timeline-chronic and Timeline-cyclical represent stronger beliefs that the condition is lasting or cyclical rather than acute in nature; higher scores on Consequences and Emotional representation represent stronger beliefs that the condition will have a negative impact on life; and higher scores on Personal control, Treatment control and Illness coherence constitute a positive belief about having control over the disease and finding treatment being effective as well as a greater personal understanding of the illness (16). The Identity component is measured by a

number of symptoms that patients are asked to evaluate in terms of whether they are a part of the illness (17 symptoms). On recommendation of the developers of the IPQ-R, Identity includes illness-specific symptoms as well as symptoms that are more general (e.g., fatigue and sleeping difficulties). Higher Identity scores indicate that a patient perceives a higher number of symptoms attributed to the illness. The IPQ-R has been found to yield adequate levels of validity and reliability (16). The Swedish version has shown good internal consistency and test-retest reliability (63). Example of questions format and responses available in the IPQ-R (Figure 3).

The Revised Illness Perception Questionnaire (IPQ-R)

Listed below are a number of symptoms (Identity scale) that you may or may not have experienced since your illness. Please indicate by ticking the appropriate box, whether you have experienced any of these symptoms since your illness, and whether you believe that these symptoms are related to you illness

	I have experienced this symptoms since my illness		This symptom is related to my illness	
	Yes	No	Yes	No
Upset stomach				

Please indicate how much you agree or disagree with the following statement about your illness by ticking the appropriate box.

Views about your illness:	Strongly disagree	Disagree	Neither agree nor disagree	Agree	Strongly agree
My illness is a serious condition					

Figure 3. Example of questions format and responses available in the IPQ-R

The Quality of Care from the Patients Perspective (QPP) was used to measure quality of care (20). The questionnaire includes four dimensions (the Medical-technical competence of the caregivers, the Physical-technical conditions of the care organization, the Identity-oriented approach and the Socio-cultural atmosphere of the care organization) and ten single items (64). The dimension *Medical-technical competence of the caregivers* includes medical care and waiting time. The dimension *Physical-technical conditions of the care organization* consists of care equipment, care room characteristics and means of communication. The dimension *Identity-oriented approach of caregivers* includes information before procedures, information after procedures, participation, responsible persons, respect, commitment, emphatic and

personal. The dimension *socio-cultural atmosphere of the care organization* is made up of general atmosphere, secluded environment, family and friends and routines. All items included in the dimensions are evaluated in two ways. First, to measure the perceived quality of experienced care respondents are asked to indicate their level of agreement on a four-point Likert scale ranging from "strongly disagree" to "strongly agree", or "not applicable", except for the physical-technical conditions for which a dichotomous response format was chosen (yes/no). Second, to measure the subjective importance the person ascribes to the various aspects of care, they are asked to indicate their level of agreement on a four-point Likert scale ranging from "of little importance" to "of very great importance". The single items included in the questionnaire measure physical and mental well-being, compliance to professional recommendations, access to health professionals and feelings about seeking care at the outpatient clinic in the future. For the purpose of this thesis, 25 items belonging to the four dimensions and eight 8 single items appropriate for outpatient care were selected. The phrasing of the items was altered to respond to patients' perception of the quality of care in general instead of referring to a specific appointment as in the original version of the questionnaire. Example of question format and responses available in the QPP (Figure 4).

Quality of Care from the Patients Perspective (QPP)

Commitment	**How I think my care really was**				
	Fully agree	Mostly agree	Partly agree	Do not agree at all	Not applicable
I had the feeling that the nurses are interested in my concerns and hassles					

	This is how important it was for me				
	Of very great importance	Of great importance	Of some importance	Of little importance	Not applicable
I had the feeling that the nurses are interested in my concerns and hassles					

Figure 4. Example of question format and responses available in the QPP.

Demographic and clinical characteristics (studies I-IV)

Participants' demographic and clinical characteristics were obtained from the national Swedish Polyposis Registry.

6.3.3 Procedures

Study I

A letter was sent to those patients at the outpatient clinic at the national SPR that fulfilled the inclusion criteria. Sixteen patients agreed to participate in the focus group interview. The participants were divided into three groups by age. Group 1 included patients aged 20-30 years (n=4, one male), group 2 patients aged 31-50 years (n=6, one male) and group 3 patients aged 51 years and older (n=6, two males). However, two participants in the youngest group declined participation with short notice so group 1 (age 20-30) finally included two participants (one male). The same moderator conducted all three focus group interviews. Interviews were carried out at the hospital after working hours; each interview lasted approximately two hours and was audio taped.

Studies II-IV

A letter was sent to those patients in the SPR that fulfilled the inclusion criteria. The letter included information about the study and contained a package of questionnaires. The letter highlighted that participation was voluntary and that non-participation would not affect a patient's care or treatment. The patients who subsequently agreed to participate were asked to complete the package of questionnaires and return it in the enclosed postage-paid envelope. Patients who did not return the questionnaire were sent a reminder after three weeks. All returned questionnaires were read to detect missing data and participants were contacted by phone for completion of data when necessary.

6.3.4 Data analyses

Study I

Data from the focus group interviews conducted in study I were transcribed verbatim and analyzed using a descriptive qualitative content analysis method (65). The analysis was performed in the following steps. (1) The transcribed material was repeatedly read to gain an overview, with words and sentences having similar meanings being identified and coded. (2) The codes were classified into categories that reflected their central content and the categories were then divided into sub-categories. (3) During the

analysis process, codes and content of the categories and sub-categories were discussed in the research group until final agreement was reached (65, 66).

Studies II-IV

Statistical calculations were performed using the Statistical Package for Social Sciences (SPSS) for Windows, version 17.0 (Studies II-III) and the IBM SPSS Statistics 19 (Study IV). Demographic and clinical characteristics were presented with means and standard deviations or proportions. P-values <0.05 were interpreted to indicate statistical significance in studies II-III. To avoid false significant findings (type 1 error) because of calculations of several analyses in study IV p-values <0.01 were applied to indicate statistical significance.

Study II

Presence and frequency of abdominal symptoms are presented with the proportions. The overall troublesomeness of symptoms was calculated by adding up the scores (possible score, 1-7) for all symptoms and dividing the sum by the number of reported symptoms. Troublesomeness of abdominal symptoms is presented with mean and range. To investigate potential differences in mean values between groups independent *t*-tests and one-way analyses of variance (ANOVAs; gender and type of surgery as independent variables) were calculated. Relationships between variables (number of symptoms, perceived troublesomeness of symptoms, age and age at first and last surgery) were studied using Pearson's correlation coefficients. Coefficients <0.29, 0.30-0.49 and >0.49 are interpreted as small, moderate and large, respectively (23).

Two three-step hierarchical multiple regression analyses were computed to determine predictors of physical and mental health (component summaries PCS and MCS of the SF-36; study II). Variables considered to influence patients' physical and mental health were entered into the model. In the first step age, gender and FAP in the family was included; in the second step age at last colorectal surgery and type of performed surgery (IRA, IPAA or ileostomy) were forced into the model; and in the third step number of symptoms and troublesomeness were forced into the model.

Study III

Descriptive statistics (means and standard deviations) were computed for components of the IPQ-R and dimensions of the SF-36. Differences in subgroups (gender, family history of FAP and adherence to endoscopic surveillance) were tested by independent *t*-tests.

To investigate internal consistency reliability Cronbach's alpha coefficients were calculated for each IPQ-R component in study III (Timeline-chronic, α=0.83; Timeline-cyclical, α=0.67; Consequences, α=0.84; Personal control, α=0.67; Treatment control, α=0.61; Emotional representation, α=0.89; Illness coherence, α=0.87). Cronbach's α in the vicinity of 0.70 or higher is considered reliable, which is why the Treatment control component was considered unreliable and therefore deleted from all analyses. Although the Cronbach's α of the component Personal control did not reach 0.70 it was considered acceptable, and therefore included in the analyses.

In study III five three-step hierarchical regression analyses were performed to determine predictors of HRQL. Results from the first study showed that living with FAP was associated with several patient concerns. Scrutinizing the identified concerns in relation to the scales of the SF-36, patient concerns were related to five of the scales: Bodily pain, General health, Vitality, Social Function and Mental health. These scales were therefore entered into the regression analyses as dependent variables. In the first step age, gender and FAP in the family were included; in the second step time since last colorectal surgery and number of symptoms were forced into the model; and in the third step six components of the IPQ-R (Timeline-chronic, Timeline-cyclical, Consequences, Personal control, Illness coherence and Emotional representation) were additionally included in the model.

Study IV

The answers to the 25 QPP items, measuring quality of care, were dichotomized into two categories: agree ("strongly agree" and "agree") and disagree ("disagree" and "strongly disagree"). The proportion of patients who agreed is presented and differences in categorical variables were tested by χ^2 tests.

Cronbach's alpha coefficients were calculated to estimate internal consistency (Identity-oriented approach, α=0.94; Medical-technical competence, α=0.75 and Socio-cultural atmosphere, α=0.70).

Logistic regression analyses were performed to examine predictors of quality of care. The three dimensions of the QPP included in study IV were dichotomized into low quality of care=0 (1st, 2nd and 3rd quartile) and high quality of care=1(4th quartile) and entered as dependent variables. The independent variables were sex (0=women, 1=men), age, time since last colorectal surgery, FAP in the family (0=no, 1=yes), organization of care (0=non-specialized care, 1=specialized care) and number of symptoms.

7 ETHICAL CONSIDERATIONS

Since World War II, several ethical guidelines have been formulated. One of the most pre-eminent documents addressing research ethics is the Declaration of Helsinki (67). The Declaration of Helsinki was adopted by The World Medical Association. The ethical principles were developed for medical research involving human subjects in the 1960s and later revised several times, most recently in 2008. The principles bind the researchers to protect life, health, dignity, integrity, right to self-determination, privacy and confidentiality (WHO). In addition, most disciplines have formulated their own code of ethics. For nurses, The American Nurses Association (ANA) nine ethical principles for the guidance of nursing research (68). Mutual for the Declaration of Helsinki and ICN is the principle of *beneficence* – do no harm – including physical, psychological and economic harm (67, 68). This thesis was preceded by a research protocol that included information about study design. Formal ethical approval of the four studies of this thesis was granted by the Regional Ethic Review Board in Stockholm (registration number: 04-241/3, 2007/1400-31/2). A letter was sent to potential participants with information about the studies. The letter also informed about the benefits of the study (e.g., that we would learn more about surgically treated individuals with FAP) in order to organize care and support based on the findings. It has been suggested that in contrast to medical research, qualitative research may be more vulnerable to ethical issues that may arise from problems uncovered in the research process (69). Therefore, participants in study I were informed about the possibility of negative psychological effects created by being involved in the focus group interviews and were therefore encouraged to call the researcher if they needed future support.

The second principle, *dignity*, includes the right to self-determination and the right of full disclosure (67, 68). To help individuals make informed decisions about participation all participants received a letter with information about the study, including a telephone number to the researcher if questions were raised. To prevent coercion the information letter emphasized that participation was voluntary and that participants could withdraw without consequences at any time during the study. To minimize the pressure of participation no reminder was sent to the participants in study I. This tactic was important because the participants in study I were recruited from the outpatient clinic affiliated to the Swedish Polyposis Registry where two of the

researchers worked. For studies II-IV, the individuals who subsequently agreed to participate were asked to complete the set of questionnaires and to return them in an enclosed pre-paid envelop. Patients who did not return the questionnaire were sent a reminder after three weeks. The third principle, *justice*, refers to participants' right of fair treatment and privacy (67, 68). In the information letter (Study I-IV) it was emphasized that non-participation or withdrawal of consent would not affect a patient's care or treatment. To provide access to research personnel at any time during the study the cell-phone number to one of the researchers (KF) was included in the information letter. Anonymity is impossible to guarantee in qualitative research; however, confidentiality was implemented by omitting the name of participants during the transcription of the interviews. Each participant was given a number for identification. Anonymity for participants in studies II-IV was also not possible because of the need for additional clinical data. To ensure confidentiality all questionnaires were provided with a number. Each patient's number code key was locked up and separated from the completed questionnaires. No names or personal numbers were entered into the data file. Furthermore, the data received from the studies are presented such that confidentiality is ensured. Interview data provided from study I are presented with quotes, but without names. For study I participants signed an informed consent before contact by the researcher (LW). For studies II-IV, no sign informed consent form was sent to potential participants. Instead, the completed questionnaires that were returned to the research group were taken as the participants' consent to participate.

8 RESULTS

8.1 CONCERNS IN LIFE AND MANAGING LIFE

The three focus group interviews were analyzed both within and across groups. Agreement on aspects of living with FAP was found between the three focus groups, whereas family planning and issues on employment were more frequently discussed in the youngest focus group. A positive interaction was found in all three focus groups but was especially salient in the two youngest focus groups, where the participants seemed to value the opportunity to discuss their personal experiences with each other. The results from the focus group interviews were revealed in two categories (*concerns in life* and *managing life*). Both categories were found to be associated with the heredity and lifelong nature of FAP, as well as to life after prophylactic colorectal surgery. In addition, *managing life* was related to acceptance of having FAP.

8.1.1 Concerns in life

Living with a hereditary disease

One aspect discussed frequently was the risk of FAP with respect to the participants' children. The discussion included concerns about the risk of children inheriting FAP and, despite this risk, a wish for having children. One young man said *"...to refrain from a child with FAP would feel like refraining from my own life-- and my life is pretty good..." Furthermore,* concerns for the health of close family members were also discussed, as one women said, *"... there was a risk that he (sibling) had developed cancer /-/ I live with this all the time..."*. Family members' deteriorating health was also a reminder of one's own risk for poorer health. One woman said, *"... I think about what is going to happen with my body in the future, especially since I have seen my brother. I am going to have an ileostomy. It's certain and just a matter of time…"*. Furthermore, being brought up in a family with FAP could result in a negative view of FAP. One woman put it in the following words: *"...my mother was the first survivor of the disease in the family; 14 relatives had died. I have my mom's bitterness against the disease. She had problems living with it…"*

Living with a lifelong disease

Concerns were related to the fact that FAP is a relatively rare disease, resulting in a lack of knowledge about the illness not only among friends but also among health care

professionals. Most participants told friends that they have FAP, but found it difficult to share deeper emotions about it because of the fear of scaring away friends. The most frequently discussed issue, was the concern related to health care professionals' lack of knowledge about FAP. One older woman with a long experience of living with FAP expressed the following: *"...they (health care professionals) don't understand what this disease is: that's my opinion /-/ they make me really annoyed. They don't understand anything /-/ no I don't think that they understand anything".* Participants also expressed that health care providers were indifferent to listen to what they had to say about their illness or to learn more about the illness, which resulted in feelings of insecurity and neglect.

Life after prophylactic surgery

Prophylactic colorectal surgery was discussed as a 'turning point', as one young man expressed it. *"... it's probably the worst thing ever /-/ when the doctor did the ward round /-/ he took a chair and closed the door /-/ and he said, 'your life is going to change from now on'. I didn't know anything about the disease then..."* Some patients described how they went from a healthy to an unhealthy state (e.g., abdominal symptoms, decreased bowel function, restrictions in food intake and reduced social life as a result of surgery). One woman expressed how she felt when visiting public toilets: *"... I panic /--/ it (the intestine) makes noises and so /--/ you can't do anything about it-- it's out of control…"* In addition, unmet needs (such as psychological support at time of surgery) were discussed. One participant noted, *"...when the surgeons were done and satisfied with the result, I felt like a victim..."* Worries about the risk of having to undergo additional surgery in the future were also frequently discussed, which was related to an unwillingness to undergo surgery, as well as the risk of not knowing how it would affect bowel function. Recurrent hospital visits had an influence on the patients, constantly reminding them of their disease: as one patient put it, *"I only feel ill once a year and that is when I have the doctor's appointment or the endoscopic examination. This is the only time I have FAP-- in between I don't...".* Moreover, the regular screening procedures, especially gastroscopy, including the laxative preparation, were described as very unpleasant.

8.1.2 Managing life

Dealing with a hereditary disease

Being raised in a family with FAP was positive for some participants. One woman put it this way, *"…my father has a stoma so we have talked about it. It (FAP) has always been in the family /-/ it has been quite easy to talk about..."* Having hopes for better treatment alternatives for their children in the future were expressed by statements such as, 'treatment for FAP will progress' and 'in the future our children may just take a tablet of medicine instead of having to undergo prophylactic colorectal surgery'.

Dealing with a lifelong disease

Thinking positive and finding positive things in life were ways of dealing with concerns associated with the lifelong disease. One patient said, *"... I usually think that I could have a disease worse than this..."* Another patient remarked, *"...yes, exactly I could have been dead..."* Finding ways to deal with the endoscopic examinations to minimize their worries varied considerably and were highly individual. For instance, some patients wanted to follow the examination on the screen, whereas others avoided looking at the screen. Some patients requested sedatives before undergoing gastroscopy, whereas others did not.

Dealing with food and toilet habits

Many patients expressed how they tried to avoid problems and pain from the intestine by such methods as 'trial and error' or by defying food restrictions and hoping for the best. The participants took and gave advice. The participants also seemed to take the opportunity to share their experiences with others with the same problems. The participants frequently discussed and shared ways of planning social activities. As one woman explained, some needed extensive planning, *"...I want to know exactly where the toilet is /-/ before I go into the cinema, I usually go two or three times to the toilet just to be sure that I can sit down during the whole movie". Other patients reported making* minor adjustments before joining social activities. Different ways of dealing with FAP at work were expressed: for instance, one young woman said, *"... I speak freely about it /-/ I have to have someone to cover up for me if I need to go to the toilet…"* Another woman said, *"...I never told anyone about it /-/ every time I had an appointment for an examination or if I felt sick I took a vacation day..."*

Learnt to live with

Most of the participants in the focus groups expressed acceptance of living with FAP. A majority of the participants felt they lived a normal life despite the illness. One participant remarked, *"... I live a normal life basically /-/ except for the examination I have to go through..."* Acceptance of having to undergo endoscopic examinations and of having to be off work during the days around the examination was also expressed. One participant with ileostomy felt independent because he no longer had to be close to a toilet.

8.2 ABDOMINAL SYMPTOMS (STUDY II)

Totally, 190 patients (91%) reported at least 1 of 21 possible symptoms, measured with the Abdominal Symptoms Questionnaire (ASQ; table 1); a mean of 7 (SD 4.61; range 1-18) symptoms was reported. Women reported a statistically significant higher number of abdominal symptoms than men (women: mean=8, SD=4.89, range 0-18; men: mean=5, SD=4.49, range 0-18), whereas no differences were found in perceived troublesomeness. Abdominal symptoms by type of colorectal surgery are presented in Table 4.

Table 4. Presence of abdominal symptoms the last three month, by type of surgery, measured with the Abdominal Symptom Questionnaire (ASQ)

	IRA (n=71)		IPAA (n=82)		Ileostomy (n=39)	
	n	**%**	**n**	**%**	**n**	**%**
Diarrhea	50	72	59	73	19	49
Borborygmi	43	62	62	77	16	41
Nighttime urge of defecation	32	46	60	74	25	64
Abdominal distension	32	46	49	60	16	41
Abdominal discomfort or pain relieved by defecation	36	52	36	44	9	23
Troublesome passage of gas	31	45	24	30	12	51
Feeling of incomplete defecation	22	32	38	47	3	8
Feeling of fullness after meals	25	37	25	31	14	36
Nausea	18	26	30	37	12	31
Retrosternal pain	23	34	23	28	13	33
Belching	20	30	25	31	12	31
Heartburn	22	32	20	25	14	36
Early satiety	20	29	29	36	7	18
Abdominal discomfort or pain on defecation	19	28	22	27	10	26
Reflux episodes	14	21	17	21	13	33
Loss of appetite	12	18	18	22	5	13
Constipation	9	13	10	12	8	21
Altering diarrhea and constipation	9	13	7	9	3	8
Vomiting	6	9	8	10	4	10
Loss of weight	6	9	7	9	2	5
Dysphagia	4	6	7	9	-	-

All 21 possible symptoms were reported by participants with IRA, IPAA and ileostomy, except for dysphagia, which was not reported by participants with ileostomy. No statistically significant differences in perceived troublesomeness or in mean number of symptoms were reported by type of prophylactic surgery.

To evaluate health status two three step hierarchical analyses were computed. The first hierarchical regression analysis was computed with the physical summary component of the SF-36 (PCS) as dependent variable. In the first step sex, age and FAP in the family were entered into the model. In step 2 time since last colorectal surgery and type of surgery (IRA, IPAA or Ileostomy) were forced into the model. Finally, in step 3 the number of symptoms and troublesomeness were forced into the model. In the final model, when the variances of the other independent variables were held constant, only age (β standardized= -0.332) and number of abdominal symptoms (β standardized= -0.465) were significant contributors to the model (R^2 change=0.217, R^2 adjusted=0.332, F=11.043).The second hierarchical regression analysis was computed with mental summary component of the SF-36 (MCS) as the dependent variable. In the first step sex, age and FAP in the family were entered into the model. In step 2 time since last colorectal surgery and type of surgery (IRA, IPAA or Ileostomy) were forced into the model. Finally, in step 3 the number of symptoms and troublesomeness were forced into the model. The final model showed that, when the variances of the other independent variables were held constant, only age (β standardized= 0,215) and number of abdominal symptoms (β standardized= -0.459) were significant contributors to the model (R^2 change=0.226, R^2 adjusted=0.329, F=10.923). The final models accounted for more than 30% of the variance of physical health and 30% of the variance of mental health.

8.3 ILLNESS PERCEPTION (STUDY III)

Overall, the results for illness perception show that the participants view FAP as a chronic illness (timeline) and possible to control to some extent (personal control). Moreover, negative impact (consequences) on life and emotional distress (emotional representation) that are due to FAP are viewed to a limited extent. High scores on the component illness coherence indicate that the participants have a good understanding of their illness (Table 5). FAP-specific symptoms were reported to be related to FAP to a high extent, indicating a strong illness identity. Furthermore, a majority of the participants reported heritage as the most important reason for having the illness. The

subgroup analysis revealed that women perceived FAP as having a more negative impact on their life (consequences) and had a stronger negative emotional response (emotional representation) to the illness than men (men mean=15.31, SD=5.42, women mean=17.97, SD=5.53, $p<0.01$; men mean=14.32, SD=5.97; women mean=16.97, SD=5.31, $p<0.01$). Another finding was that participants with a history of FAP in the family had a greater understanding (illness coherence) of the illness and perceived a less negative impact (consequences) on life compared with patients without a history of FAP in the family (FAP in the family mean=20.62, SD=4.03, without FAP in the family mean=17.75, SD=4.16, $p<0.001$; FAP in the family mean=16.20, SD=5.54, without FAP in the family mean=19.28, SD=5.38, $p<0.01$). Individuals who were adherent to endoscopic surveillance perceived the illness to be chronic (timeline-chronic) to a higher extent and perceived more negative emotions (emotional representation) to the illness than participants' who were non-adherent to endoscopic surveillance (adherent mean=27.29, SD=3.32, non-adherent mean=25.52, SD=4.92, $p<0.05$; adherent mean=16.14, SD=5.67, non-adherent mean=12.90, SD=5.39, $p<0.01$).

Table 5. Illness perception questionnaire, measured with the Revised Illness Perception Questionnaire (IPQ-R)

	mean	**SD**
Timeline - chronic[f]	27.04	3.62
Timeline - cyclical[g]	10.52	3.37
Consequences[h]	16.79	5.63
Personal control[i]	16.69	4.48
Illness coherence[j]	20.07	4.20
Emotional representation[k]	15.69	5.73

Meaning of higher score:
[f]*Timeline (chronic)* - stronger beliefs that the condition is lasting rather than acute in nature; possible score 6-3
[g]*Timeline (cyclical)*- stronger beliefs that the condition is cyclical rather than acute in nature; possible score 4-20
[h]*Consequences* - stronger beliefs that the illness will have a negative impact on the person's life; possible score 6-30
[i]*Personal control*- positive beliefs about personal control; possible score 6-30
[j]*Illness coherence* – a greater perceived personal understanding of the illness; possible score 5-25
[k]*Emotional representation* - stronger negative emotional responses to the illness; possible core 6-30

8.4 HEALTH-RELATED QUALIT Y OF LIFE (STUDY III)

HRQL, measured by the eight subscales of the SF-36, showed that women perceived a worse HRQL than men in five of the eight subscales (Physical function, Role physical, Bodily pain, General health and Vitality). To investigate predictors of HRQL five three step hierarchical analyses were computed with the SF-36 subscales (Bodily Pain, General health, Vitality, Social function and Mental health) as dependent variables (Table 6). When the effect of gender, age and FAP in the family was already accounted for (first step), the number of abdominal symptoms (second step) had a statistically significant effect on all subscales. In the third step the addition of the components of the IPQ-R contributed to the models in all subscales except for Mental health. The final models accounted for 33 to 58% of the variance in HRQL.

Table 6. Hierarchical multiple regression analyses with the subscores of the SF-36 as dependent variables (n=190)

	Bodily pain		**General health**		**Vitality**		**Social function**		**Mental health**	
	R^2 change	R^2 adjusted	R^2 change	R^2 adjusted	R^2 change	R^2 adjusted	R^2 change	R^2 adjusted	R^2 change	R^2 adjusted
Step										
1[c]	0.075**	0.060	0.061**	0.046	0.065**	0.050	0.052*	0.037	0.062**	0.047
2[d]	0.215***	0.271	0.362***	0.408	0.343***	0.391	0.297***	0.331	0.254***	0.298
3[e]	0.074	0.324	0.182***	0.581	0.074**	0.450	0.109***	0.425	0.111***	0.392

*p<0.05, **p<0.01, p<0.001, The sub scores of the SF-36 range from 0 to 100 with higher scores indicating better health status
[C]Predictors: gender, age, FAP in the family; [d]Predictors: time since last surgery, number of symptoms [e]Predictors: IPQ-R components: timeline-chronic, timeline-cyclical, consequences, personal control, treatment control, illness coherence, emotional representation

8.5 QUALITY OF CARE (STUDY IV)

Overall quality of care was rated high for all three dimensions (Identity-oriented approach: median=4, IQR 2-4; Medical-technical competence: median=4, IQR 3-4; Socio-cultural atmosphere: median=4, IQR 2-4). Individuals receiving specialized care reported statistically significant higher agreement, reflecting better care, on 18 of the 25 statements included in the three dimensions of the QPP. Accessibility was reported to be easy or very easy to a higher extent by individuals receiving specialized care compared with individuals receiving non-specialized care (specialized care 60% vs. non-specialized care 40%, $p<0.01$). Furthermore, in comparison with patients receiving non-specialized care, a lower proportion of individuals receiving specialized care hesitated to revisit the clinic (specialized care 4% vs. non-specialized care 26%, $p<0.001$).

Logistic regression analyses were carried out with the three dimensions (Identity-oriented approach, Medical-technical competence and Socio-cultural atmosphere) of the QPP serving as dependent variables. The models showed that individuals receiving specialized care and those with a lower number of abdominal symptoms were more likely to report good quality of care than individuals receiving non-specialized care and those with a higher number of abdominal symptoms. Odds ratios (ORs; the probability of an event) for organization of care ranged from 14.332 to 8.049 while ORs for number of symptoms ranged from 0.871 to 0.898 (Table 7).

Table 7. Logistic regression analyses with the 3 dimensions in the Quality of Care from the Patients Perspective (QPP) as dependant variables[a] (n=122-205)

Dimensions		Sex[b]	Age	History of FAP in the family[c]	Time since last colorectal surgery	Organization of care[d]	Number of symptoms
Identity-oriented approach	**OR**	2.424	1.013	1.875	0.995	14.332**	0.871*
	99% C.I	0.880-6.674	0.973-1.055	0.547-6.433	0.943-1.051	2.536-81.005	0.782-0.971
Medical-technical competence	**OR**	1.346	1.008	0.962	1.008	8.049**	0.898*
	99% C.I	0.547-3.308	0.974-1.043	0.313-2.955	0.959-1.060	2.345-27.628	0.812-0.993
Socio-cultural atmosphere	**OR**	1.044	0.999	1.484	0.989	9.764**	0.844*
	99% C.I	0.366-2.976	0.959-1.040	0.437-5.040	0.936-1.045	1.754-54.361	0.753-0.947

[a]low quality of care=0, high quality of care=1, [b]men=0, women=1; [c]FAP in the family yes=0, no=1; [d]specialized care=0, non-specialized care=1

*p<0.01, **p<0.001

9 DISCUSSION

9.1 GENERAL DISCUSSION OF THE FINDINGS

The results of the four studies included in this thesis reveal that surgically treated individuals with FAP perceive their lives to be influenced by having FAP in several ways. Unspecific abdominal symptoms have not previously been investigated to the same extent as they have in this thesis. A majority of the participant's perceived abdominal symptoms and in contrast to other studies (11) that was independent on type of surgery. Interestingly, when calculating t-tests to compare means of SF-36 scales between the individuals with FAP and published norm data for the general Swedish population (61) individuals with FAP appear to have a diminished physical health (Role function, Bodily pain, General health and Social function; data not shown). This corroborates to the results showing a high symptom burden and need of health care in this group found in study I and II However, the norm values for the Swedish population are gathered more than ten years ago why this should be interpreted with caution. An important finding of this thesis is the influence of abdominal symptoms on HRQL. HRQL worsened with increasing number of abdominal symptoms. The focus group interviews gave insight into how social life could be influenced by a person's symptom (e.g., visiting toilets outside the home was associated with a feeling of lack of control because of the unpredictable passage of gas). Further, the qualitative data revealed the impact on life caused by symptom clusters (10), one individual pointed out that it was sometimes difficult to figure out what was wrong. She called days like that "belly days", and that was when her stomach was upset. The results indicate that an effective management of symptoms would improve HRQL, in line with a study of individuals with ulcerative colitis who receive similar treatment to patients with FAP (70).

The prophylactic surgery was perceived as a turning point from which individuals went from a healthy to an unhealthy state and felt a lack of control. In contrast to other studies (47, 55) that was independent of type of surgery and age at time of surgery. Despite the fact that the prophylactic surgery may be seen as traumatic and the reason for a wide range of abdominal symptoms, the illness perception of individuals, revealed from both quantitative and qualitative data, in this thesis indicates that they, in general, adjust well to the illness. However, the illness perception of individuals without FAP in

the family showed that they had a poorer understanding of the illness and a perception of more negative consequences of the illness compared to individuals with FAP in the family. Although, the results from the focus group interviews revealed a more complex picture of the effect of the illness on individuals who have family members with FAP. The results showed that having family members with FAP was associated with concerns for the health of family members. Further, when family members had additional operations or their illness deteriorated, increasing worries about their own health were perceived. The use of a wide range of self-care strategies to minimize the effect of the illness on life, found in the focus group interviews, may cause the health care providers to overlook symptoms and concerns of their patients. Individuals with chronic illnesses may not always discuss with health care professionals problems about their everyday life, nor what kinds of self-care actions they use (6). Individuals who are newly diagnosed may need extra attention in that self-care decisions can be overwhelming in the initial phase of an illness, but over time demand less attention (4).

This thesis revealed some differences between men and women. Women reported more symptoms than men. With regard to illness perception women had more negative emotions and perceived more negative consequences to the illness than men. However, surgically treated women with FAP may not differ to women from the general population since women report abdominal symptoms (71) and health complaints to a higher extent than men (72). Non-adherence was found to associate with lower emotional responses to the illness and a perception of FAP as less chronic. Such results, however, are in contrast to a study showing that higher emotional responses and low personal control were associated with non-adherence in individuals with hypertension (73). The differences in results are hard to explain since the reason for non-adherence in the present thesis was not explored. However, it is well-known that worry and anxiety are motivators of illness behavior (74). One possible contributing reason for non-adherence may be related to organization of care. Individuals perceiving high quality of care were more likely to receive specialized care and present with a lower number of abdominal symptoms. Specialized care is provided with the aim of gaining and maintaining positive relations with individuals, important for building partnerships (75) and for an individual's participation (76, 77) and adherence to professional recommendations (78). Knowledge of the organizational structures and priorities of the clinics outside the Swedish Polyposis Registry is gained from medical records and may therefore not be comprehensive. Nonetheless, according to the medical records, it is

common that several physicians are involved in the care of an individual, and that in general nursing care is lacking. It also seems that although several individuals with FAP are affiliated to the same hospital/clinic, they often receive care from different physicians. This situation may result in a lack of combined knowledge, which is especially important when caring for individuals with such a rare disease.

The results of the studies in this thesis indicate that surgically treated individuals with FAP would benefit from a care that focuses on the support of the individual's self-care (6). To improve HRQL the management of abdominal symptoms is vital, as negative consequences on life perceived by the individual. As the discipline of nursing is based on a holistic approach, nurses are suggested to be especially important when caring for chronically ill individuals. It is important that interventions fit in with an individual's day-to-day life, and an approach not limited to medical characteristics (e.g., type of surgery) (6). In the context of FAP having diverse interaction approaches (e.g., support groups, follow-up programs) (6), patient education (individual or group), education for family members and interventions over the internet and phone would be beneficial (6, 79). However, one obstacle previously discussed which also is transmittable to health care in the Swedish context, is the difficulty of getting reimbursement for non-visiting methods of interaction (79). In the Stockholm county council reimbursement is (almost) only provided for face-to-face physician-patient interactions that includes some form of medical treatment. This restriction is highly problematic when caring (e.g., supporting self-care) for individuals with chronic diseases, such as FAP.

9.2 METHODOLOGICAL CONSIDERATIONS

9.2.1 study design

A major strength of the thesis is the use of both a qualitative and quantitative approach, which allowed for insight into how individuals with FAP perceive life after surgery, from different angles. However, the cross-sectional design of the studies precludes the assessment of causal relationships (80). Furthermore, because no control group was used, the information cannot be considered conclusive.

9.2.2 Focus group interviews

The rich data source of the focus group interviews indicates the appropriateness of the method of data collection. Classifying the participants into groups based on age seemed

to facilitate the discussion since some of the areas (such as family planning and career issues) were more frequently discussed in the youngest group. Participants discussed areas not included in the interview guide, such as individual's relationship to health care professionals and ways of managing everyday life problems. The easiness of discussing sensitive topics in focus groups has been questioned (81). Concerns about developing cancer in the future were not mentioned in the present study. One possible explanation could be that the participants perceived the risk for future cancer to be too sensitive to bring up in a group session, which is one reason why individual interviews may have been a more appropriate method. Still, it is also possible that individuals do not view FAP as a cancer disease since the majority (79%) of the participants in the focus group interviews never had cancer.

In qualitative studies methodological considerations refer to trustworthiness defined by terms such as credibility, dependability and transferability (66). Credibility pertains to the focus of the research and the appropriateness of data collection, the amount of data gathered and analysis of the data, as well as context selection (66). In the current study the participants represented all ages, both men and women participated and they had experience of living with symptoms as a result of different surgical methods, which contributes to a rich variation of the phenomena under study (81). In comparison with other qualitative methods, interaction is a unique part of the focus group method. It has been argued that focus group interviews may have an intervention effect rather than being solely a research method. However, the loquaciousness of the focus group participants during the interviews was a result of the positive attitudes toward meeting other persons in the same situation. This provided rich data from all three focus groups and greatly helped in answering the research questions. The literature usually recommends 4-8 participants in a focus group (81). The low number (n=2) of participants in the youngest focus group was due to the last minute withdrawal of participants. The decision to carry on with the interview was made because the two participants had invested their time in coming. Since the data was considered sufficient and the content from all three focus groups were analyzed together, no further attempts to gather young persons with FAP to an additional focus group were made. Another critical issue for achieving credibility is the quality of the data analysis (66). The first author in the study (KF), who conducted the early stage analysis of the data, has extensive experience in clinical practise of patients with FAP. Henceforth, during the analysis process, codes, subcategories and categories were discussed among the co-

authors (LW, CP) until a final agreement was reached (65, 66). Whether pre-understanding is an advantage or disadvantage in qualitative research is not clear but some consider pre-understanding to be an obstacle for an objective analysis (82). Since the main researcher who analyzed the interviews had extensive knowledge in the participating patients' illness, her pre-understanding and its possible influence on the analysis process were continuously discussed among the co-authors. One co-author (CP) involved in the analysis process came from outside the research group and had no experience of FAP. Furthermore, one of the co-researchers (LW) had many years of research experience from the perspective of patients with cancer within the field of nursing sciences but had no previous knowledge in FAP. Dependability deals with the degree to which data change during data collection or during the analysis process (66). To prevent alterations in data collection procedures and preserve data integrity the same moderator (LW), collected the data from all three focus groups. In addition, the use of an interview guide prevents inconsistencies when questioning participants (66). Transferability refers to what extent the findings can be transferred to other settings (66). In what way the results of the focus group interviews can be generalized to other settings is uncertain. The findings reflect the situation of patients at a specialized clinic in Sweden and may not be representative of the FAP population as a whole. This must be seen as a limitation of the study. Another point to consider is that there were more women than men participating and the mean age of the participants was relatively high. Nevertheless, the findings may partly be transferable to other settings in that the study participants, in comparison with the Swedish FAP population, did not differ on several characteristics (e.g., time since diagnose, type of surgery, desmoid tumours and FAP-related cancer).

9.2.3 Validity and reliability

Validity refers to what extent a measure reflects the latent construct it is intended to measure and to which extent the results can be reproduced (80). A major strength of studies with a quantitative approach in this thesis is the unique representative cohort of surgically treated individuals with FAP. The sample was recruited from the national Swedish Polyposis Registry, which is considered to cover nearly the entire FAP population in Sweden. The response rate was 76%; no differences in demographic and clinical characteristics between responders and non-responders were found, why the risk of selection bias was limited. Because of the high response rate regression analyses that allow controlling for confounders (factors that associate with outcome) could be

used (Study II-IV). To increase the interpretation of the results major efforts were made to detect and complete missing data. Given the high response rate and the sample recruiting mentioned above, the generalizability to the Swedish FAP population who have undergone prophylactic colorectal surgery must be considered high.

The use of a questionnaire not previously used to investigate abdominal symptoms in this population (i.e. the ASQ) could be a threat to the validity of the study. The results of study II emphasize the appropriateness of using the ASQ in that the abdominal symptoms included in the questionnaire were frequently reported by the participants, indicating their importance in this context (23).

Reliability refers to whether an instrument measures the concept in a consistent manner (80). The questionnaire used to assess illness perception has been employed in individuals with different disorders; however, its use in a genetic disease context is limited. The reliability of the component Treatment control was not acceptable and therefore deleted from the analyses (23). That may be a result of the phrasing of the items, it has been suggested that questions about treatment effectiveness in controlling, improving and curing an inherited disease may seem irrelevant (83). This was further emphasized by a study of two nurses who tested the IPQ-R in individuals with two chronic bowel diseases (Ulcerative Colitis and Crohn's disease) using a think-aloud strategy (84). The results of that study confirm the problem of measuring treatment control by asking about treatment effectiveness in curing a chronic disease.

Measuring HRQL in surgically treated individuals with FAP with a generic rather than disease-specific questionnaire may not capture all aspects of HRQL. However, it has been suggested that measuring quality of life with both a generic and a disease-specific questionnaire are relevant (85). The results from this thesis indicate the relevance of including aspects that have a negative impact on life in individuals with FAP. However, no such questionnaire exists today. For future studies, a more individualized approach in which individuals define what quality of life means to them in relation to goals and expectations may be beneficial.

One limitation of study IV in its measure of quality of care is item selection, i.e. 25 items of 47 from three of four dimensions were included in the study. The item selection refers to the aspects of efficiency. To reduce the amount of time for

individuals to answer the questions included in the questionnaire package the most relevant items were selected. Measuring quality of care is a well-known methodological difficulty in that patients in general report high levels of perceived quality of care independent of care received and questionnaire used (86, 87), resulting in skewed data and ceiling effects. Such effects were also found in study IV. Well-validated questionnaires measuring quality of care or satisfaction in the outpatient context (and in a broader sense than the physician-patient dyad) are difficult to find. One advantage of the QPP is the items regarding quality of care delivered by nurses, which must be considered important in the evaluation of quality of care because they are highly involved in the care of individuals. To avoid the risk of a type 1 error (false positive result) because of the large number of analyses carried out in study IV the statistical significance level used was lowered (80).

9.3 IMPLICATIONS FOR CARE

In the context of FAP, support in self-care should address patient concerns in several areas, including the management of abdominal symptoms in accordance with the individuals' own strategies in managing those concerns. Individuals may benefit from pre- and post-surgery support. Further, the laxative procedure and concerns regarding endoscopic examinations should be addressed and interventions should be discussed with the individual. However, when supporting self-care, the most important factor to remember is that the individuals are the expert regarding their unique needs. Care for surgically treated individuals with FAP should be provided by a team specialized in FAP including genetic counselors, physicians, surgeons, welfare officers and dieticians, preferably coordinated by a nurse. The coordinating nurse should be specialized in the care of chronically ill individuals, including support of self-care by building partnership with each individual, promoting positive thoughts about the individual's self, facilitating the individuals control over their life situation, building on the individuals' capabilities and establishing mutual health-directed goals. Support self-care implies addressing the needs of an individual: simply asking individuals about what consequences the illness will have on their life may be the best way to individualize interventions. Because of the rareness of FAP, care should be centralized to a few university hospitals in the country.

The Swedish Polyposis Registry brings patients and relatives together for special education days once a year with the aim of improving FAP knowledge and

disseminating the latest guidelines and research in the field. Although participants are from throughout the country, not all individuals have the possibility to go to Stockholm to attend this education. More innovative ways of disseminating guidelines and research, as well as bringing individuals together may be beneficial. Using the Internet is one means of narrowing the distance between patients. A chat group for patients with FAP has been developed by a patient support group and patients have created a FAP group on Facebook. Unfortunately, at present, the SPR does not provide a website that allows networking and 'chat groups'. In addition to other interactive websites, it would be beneficial if the Swedish Polyposis Registry could provide a platform for interactions because the website includes updated information about FAP and treatment guidelines, as well as information about the clinic affiliated to the Swedish Polyposis Registry.

10 CONCLUSION

The findings reveal that life of surgically treated individuals with FAP is influenced in several ways including issues about hereditary, the life-long nature of the illness and abdominal symptoms. A high number of abdominal symptoms was reported; this had a substantial impact on HRQL and self-reported quality of care. Patients undergoing follow-up at a specialized clinic rated quality of care better than those cared for at other clinics. Despite concerns related to the illness, individuals with FAP appear to get along with life quite well.

11 FUTURE PERSPECTIVE

Future research could build on the results from this thesis by investigating the impact of symptom clusters, the relationships to family and friends and economic concerns, all of which would help us gain valuable information about FAP and its influence on the patient's daily life. Moreover, future research is recommended to study the effects of endoscopic surveillance on the individual's life, particularly the impact that laxative procedures and the examination process has on the individual's well-being. Future research pertaining to quality of care is recommended by investigating the impact of inpatient care, with particular focus on days of inpatient care and reasons for inpatient care. Knowledge about anticipatory factors of inpatient care is important when supporting self-care in order to prevent inpatient care and reduce the impact on life.

12 SAMMANFATTNING PÅ SVENSKA

Familjär adenomatös polypos (FAP) är en relativt ovanlig, ärftlig sjukdom som medför en mycket hög risk för cancer. I preventivt syfte genomgår alla personer med FAP kolektomi (borttagande av tjocktarmen) samt följer ett livslångt endoskopisk undersökningsprogram.

Det övergripande syftet med avhandlingen var att få en djupare förståelse för hur personer med FAP, efter kirurgi, uppfattar att livet är påverkat, för att utifrån resultaten utveckla och förbättra vården för dessa personer.

I den första studien genomfördes fokusgruppintervjuer med personer som genomgått kirurgi, i åldrarna 18-75 år. Studierna II-IV är tvärsnittsstudier baserade på frågeformulärsdata. I Studie II undersöktes gastrointestinala symtom samt hälsostatus med frågeformulären Abdominal Symptom Questionnaire (ASQ) och Medical Outcomes Study Short Form 36 Health Survey (SF-36). I Studie III, undersöktes sjukdomsuppfattning och hälsorelaterad livskvalitet med frågeformulären Revised Illness Perception Questionnaire (IPQ-R) och SF-36. I studie IV undersöktes slutligen vårdkvalitet med frågeformuläret KUPP- Kvalitet ur patientens perspektiv.

Resultatet från fokusgruppintervjuerna visar att personer med FAP som genomgått förebyggande kirurgi anser att livet är påverkat på många sätt. Påverkan på livet var relaterat till att FAP är en ärftlig och livslång sjukdom, samt till gastrointestinala symtom och en försämrad mag-tarmfunktion efter kirurgi. Vidare visar resultatet att en mängd olika strategier för att minska denna påverkan används. Resultat från studien som undersökte gastrointestinala symtom visade att 91 % av deltagarna hade haft minst ett gastrointestinalt symptom de senaste tre månaderna. Ett medelvärde på 7 (SD 4.61; spridning 1-18) symtom rapporterades. Vidare visade studien att ett ökat antal gastrointestinala symtom predicerar en sämre fysisk och mental hälsa. Resultat från studien som studerade sjukdomsuppfattning visade att FAP anses vara en kronisk sjukdom som inte är så lätt att kontrollera. Vidare visade studien att negativa konsekvenser av sjukdomen samt ett ökat antal gastrointestinala symtom predicerar sämre hälsorelaterad livskvalitet. Studien som studerade vårdkvalitet visade att personer som erhåller specialistvård och de med ett lägre antal gastrointestinala symtom hade större sannolikhet att vara nöjda med erhållen vård.

Sammantaget så visar studierna i avhandlingen på betydelsen av stöd gällande egenvård av gastrointestinala symtom samt de negativa konsekvenser som sjukdomen medför. Genom att lindra och bota symtom samt genom att minska de negativa konsekvenserna av FAP, kan hälsorelaterad livskvalitet hos personer med FAP förbättras. Vidare så bör vården för personer med FAP ges av en grupp, specialiserade på FAP, bestående av olika professioner och som med fördel kordineras av en sjuksköterska. Omhändertagandet bör genomsyras av en holistisk vård, vilket innebär att det är den enskilda individen som styr vad som är viktigt för just honom eller henne.

13 TILLKÄNNNAGIVANDEN

Först så vill jag tacka er som deltog i fokusgrupperna för att ni så generöst delade med er av era erfarenheter av att leva med FAP. Ett lika stort tack till er som orkade igenom de 150 frågorna i frågeformuläret, utan er hade denna avhandling inte blivit av.

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14 REFERENCES

1. Thorne S, Paterson B, Acorn S, Canam C, Joachim G, Jillings C. Chronic illness experience: Insights from a metastudy. Qual Health Res. 2002;12(4):437-52.
2. Curtin M, Lubkin I. What is chronicity? In I. Lubkin, editors. Chronic illness: Impact and Interventions 3rd ed. Sudbury: Jones & Barlett; 1995.
3. Thorne S. The science of meaning in chronic illness. Int J Nurs Stud. 1999;36:397-404.
4. Thorne S, Pateson B, Rusell C. The structure of everyday self-care decision making in chronic illness. Qual Health Res. 2003;13(10):1337-52.
5. Larsen PD, Lubkin IM. Chronic illness. Impact and intervention. 7th ed. Sudbury: Jones & Bartlett; 2009.
6. Heartz JE. Self-care. In: Larsen PD, Lubkin IM, editors. Chronic illness. Impact and intervention. 7th ed. Sudbury: Jones & Bartlett; 2009. p. 299-340.
7. Rhodes VA, Watson PM. Symptom distress-the concept: past and present. Semin Oncol Nurs. 1987;3(4):242-7.
8. Oxford Advanced Learner's Dictionary of Current English. 6th ed. Oxford: Oxford University Press; 2000. Apraxia; p. 1319.
9. Kugelman R. Pain as a symptom, pain as a sign. Health. 2003;7(1):29-50.
10. Armstrong TS. Symptom Experience: A Concept Analysis. Oncol Nurs Forum. 2003;30(4):601-6.
11. Aziz O, Athanasiou T, Fazio VW, Nicholls RJ, Darzi AW, Church J, et al. Meta-analysis of observational studies of ileorectal versus ileal pouch-anal anastomosis for familial adenomatous polyposis. Br J Surg. 2006;93(4):407-17.
12. Meyer D, Leventhal H, Gutmann M. Common-sense models of illness: the example of hypertension. Health Psychol. 1985;4(2):115-35.
13. Diefenbach MA, Leventhal H. The Common-Sense Model of Illness Representation: Theoretical and Practical Considerations. J Soc Distress Homeless. 1996;5(1):11-38.
14. Leventhal H, Leventhal EA, Contrada RJ. Self-regulation, health, and behavior: A perceptual-cognitive approach. Psychol Health. 1998;13(4):717-33.
15. Cameron LD. Conceptualizing and assessing risk perceptions: A self-regulatory perspective. Conceptualizing and measuring risk perception workshop.

Washington DC. 2003:p1-12 [Internet]. Available from: http://cancercontrol.cancer.gov/brp/presentations/cameron.pdf.

16. Moss-Morris R, Weinman J, Petrie KJ, Horne R, Cameron LD, Buick D. The revised illness perception questionnaire (IPQ-R). Psychol Health. 2002;17(1):1-16.
17. Thorne S. Patient-provider communication in chronic illness. A health promotion window of opportunity. Fam Community Health. 2005;29(1S):s4-11.
18. Dancer S, Courtney M. Improving diabetes patient outcomes: Framing research into the chronic care model. J Am Acad Nurse Pract. 2009;22(11):580-5.
19. Wilde B, Starrin B, Larsson G, Larsson M. Quality of care from a patients' perspective. A Grounded Theory Study. Scand J Caring Sci. 1993;7:113-20.
20. Larsson G, Wilde Larsson B, Munck IME. Refinement of the questionnaire 'quality of care from the patient's perspective' using structural equation modeling. Scand. J. Caring Sci. 1998;12(2):111-118.
21. WHO. World Health Organization. Concepts of health and disease. Reading, Massachusetts: Addison-Weasly; 1947.
22. Schirm V. Quality of life. In: Larsen PD, Lubkin IM, editors. Chronic illness. Impact and intervention. 7th ed. Sudbury: Jones & Bartlett; 2009. p. 139-57.
23. Fayers PM, Machin D. Quality of life. The assessment, analysis and interpretation of patient-reported outcomes. 2nd ed. Chichester; Hoboken, NJ: J. Wiley; 2007.
24. Ware JE, Sherbourne CD. The MOS 36-item short-form health survey (SF-36) .1. Conceptual-framework and item selection. Med. Care. Jun 1992;30(6):473-83.
25. Alm T. Hereditary Adenomas of the colon and rectum in Sweden [PhD thesis]. The Department of Gastroenterology, St Erik's Hospital: Stockholm; 1974.
26. Dukes CE. The control of precancerous conditions of the colon and rectum. Canadian Medical Association Journal. 1964;90(7):630-5.
27. Heald RJ, Bussey HJ. Clinical experiences at St Marks' synchronous cancers of the colon and rectum. Dis Colon Rectum. 1975;18(1):6-10.
28. Björk J. Studies on familial adenomatous polyposis. [PhD thesis]. Department of Medicine: Stockholm; 2000.

29. Björk J, Akerbrant H, Iselius L, Alm T, Hultcrantz R. Epidemiology of familial adenomatous polyposis in Sweden: changes over time and differences in phenotype between males and females. Scand J Gastroenterolog. 1999;34(12):1230-5.
30. Nishischo I, Nakamura Y, Mioyoshi Y, Miki Y, Ando H, Horii A, et al. Mutations of chromosome 5q21genes in FAP and colorectal cancer patients. Science. 1991;9:253(5020):665-9.
31. Al-Tassan N, Chmiel NH, Maynard J, Fleming N, Livingston AL, Williams GT. Inherited variants of MYH associated with somatic G:C-T:A mutations in colorectal tumors. Nature Genetics. 2002;30:227-30.
32. Vasen HF, Moslein G, Alonso A, Aretz S, Bernstein I, Bertario L, et al. Guidelines for the clinical management of familial adenomatous polyposis (FAP). Gut. 2008;57(5):704-13.
33. Cruz-Correa M, Giardello FM. Familial adenomatous polyposis. Gastrointest endosc. 2003;58(6):885-8.
34. Björk JA, Akerbrant HI, Iselius LE, Hultcrantz RW. Risk factors for rectal cancer morbidity and mortality in patients with familial adenomatous polyposis after colectomy and ileorectal anastomosis. Dis Colon Rectum. 2000;43(12):1719-25.
35. Björk J, Akerbrant H, Iselius L, Bergman A, Engwall Y, Wahlström J, et al. Periampullary adenomas and adenocarcinomas in familial adenomatous polyposis: cumulative risks and APC gene mutations. Gastroenterology. 2001;121(5):1127-35.
36. Kanter-Smoler G, Björk J, Fritzell K, Engwall Y, Hallberg B, Karlsson G, et al. Novel Findings in Swedish Patients with MYH-Associated Polyposis: Mutation Detection and Clinical Characterization. Clin Gastroenterol Hepatol. 2006;4:499-506.
37. Trepanier A, Ahrens M, McKinnon W, Peters J, Stopfer J, Campbell Grumet S, et al. Genetic cancer risk assessment and counseling: Recommendations of the national society of genetic counseling. J Genet Couns. 2004;13(2):83-114.
38. Duncan RE, Gilliam L, Savulescu J, Williamson R, Rogers JG, Delatycki MB. "You're one of us now": Young people describe their experiences of predictive genetic testing for huntington disease (HD) and familial denomatous polyposis (FAP). Am J Med Genet C Semin Med Genet. 2008;148C:47-55.

39. Duncan RE, Gillam L, Savulescu J, Williamson R, Rogers JG, Delatycki MB. The challenge of developmentally appropriate care: predictive genetic testing in young people for familial adenomatous polyposis. Fam Cancer. 2010;9(1):27-35.
40. Andrews L, Mireskandari S, Jessen J, Thewes B, Solomon M, Macrae F, et al. Impact of familial adenomatous polyposis on young adults: attitudes toward genetic testing, support, and information needs. Genet Med. 2006;8(11):697-703.
41. Douma KF, Aaronson NK, Vasen HF, Verhoef S, Gundy CM, Bleiker EM. Attitudes toward genetic testing in childhood and reproductive decision-making for familial adenomatous polyposis. Eur J Hum Genet. 2010;18(2):186-93.
42. Björk J, Akerbrant H, Iselius L, Svenberg T, Öresland T, Påhlman L, et al. Outcome of primary and secondary ileal pouch-anal anastomosis and ileorectal anastomosis in patients with familial adenomatous polyposis. Dis Colon Rectum. 2001;44(7):984-92.
43. Seidel SA, Newman M, Sharp KW. Ileoanal pouch versus ileostomy: is there a difference in quality of life? Am Surg. 2000;66(6):540-6; discussion 546-7.
44. Mills SJ, Chapman PD, Burn J, Gunn A. Endoscopic screening and surgery for familial adenomatous polyposis: dangerous delays. Br J Surg. 1997;84:71-7.
45. Douma KF, Bleiker EMA, Aaronson NK, Cats A, Gerritsma MA, Gundy CM, et al. Long-term compliance with endoscopic surveillance advice for familial adenomatous polyposis (FAP). Colorectal Dis. 2010;12(12):1198-207. doi: 10.1111/j.1463-1318.2009.02008.x
46. Kinney AY, Hicken B, Simonsen SE, Venne V, Lowstuter K, Balzotti J, et al. Colorectal cancer surveillance behaviors among members of typical and attenuated FAP families. Am J Gastroenterol. 2007;102(1):153-62.
47. Osterfeld N, Kadmon M, Brechtel A, Keller M. Preoperative and postoperative quality of life in patients with familial adenomatous polyposis. Dis Colon Rectum. 2008;51(9):1324-30.
48. Wuthrich P, Gervaz P, Ambrosetti P, Soravia C, Morel P. Functional outcome and quality of life after restorative proctocolectomy and ileo-anal pouch anastomosis. Swiss Medical Weekly. 2009;139(13-14):193-7.
49. Ko CY, Rusin LC, Schoetz DJ Jr, Moreau L, Coller JA, Murray JJ, et al. Does better functional result equate with better quality of life? - Implications for

surgical treatment in familial adenomatous polyposis. Dis Colon Rectum. 2000;43(6):829-35; discussion 835-7.

50. Ko CY, Rusin LC, Schoetz DJ, Coller JA, Murray JJ, Roberts PL, et al. Using quality of life scores to help determine treatment: is restoring bowel continuity better than an ostomy? Colorectal Dis. 2002;4(1):41-7.
51. Müller MW, Dahmen R, Köninger J, Michalski CW, Hinz U, Hartel M, et al. Is there an advantage in performing a pancreas-preserving total duodenectomy in duodenal adenomatosis? Am J Surg. 2008;195(6):741-8.
52. Olsen KÖ, Juul S, Bulow S, Järvinen HJ, Bakkas A, Björk J, et al. Female fecundity before and after operation for familial adenomatous polyposis. Br J Surg 2003;90:227-31.
53. Ko CY, Rusin LC, Schoetz DJ Jr, Moreau L, Coller JC, Murray JJ, et al. Long-term outcomes of the ileal pouch anal anastomosis: The association of bowel function and quality of life 5 years after surgery. J Surg Res. 2001;15:98(2):102-7.
54. Douma KF, Bleiker EM, Vasen HF, Gundy CM, Aaronson NK. Quality of life and consequences of daily life of familial adenomatous polyposis (FAP) family members. Colorectal Dis. 2010;5. [Epub ahead of print]
55. Andrews L, Mireskandari S, Jessen J, Thewes B, Solomon M, Macrae F, Meiser B. Impact of familial adenomatous polyposis on young adults: quality of life outcomes. Dis Colon Rectum. 2007;50(9):1306-115.
56. Mireskandari S, Sangster J, Meiser B, Thewes B, Groombridge C, Spigelman A, etal. Psychosocial Impact of Familial Adenomatous Polyposis on Young Adults: A Qualitative Study. J Genet Couns. 2009;18(5):409-17.
57. Esplen MJ, Berk T, Butler K, Gallinger S, Cohen Z, Trinkhaus M. Quality of life in adults diagnosed with familial adenomatous polyposis and desmoid tumor. Dis Colon Rectum. 2004;47(5):687-95; discussion 695-6.
58. Miller HH, Bauman LJ, Friedman DR, DeCosse JJ. Psychosocial adjustment of familial polyposis patients and participation in a chemoprevention trial. Int J Psychiatry Med. 1986;16(3):211-30.
59. Douma KF, Aaronson NK, Vasen HF, Gerritsma MA, Gundy CM, Janssen EPC, et al. Psychological distress and use of psychosocial support in familial adenomatous polyposis. Psychooncology. 2010;19(3):289-98.

60. Agreus L, Svärdsudd K, Nyren O, Tibblin G. Reproducibility and validity of a postal questionnaire. The abdominal symptom study. Scand J Prim Health Care. 1993;11(4):252-62.
61. Sullivan MK, J. Taft C. Svensk manual och tolkningsguide, 2:a upplagan (Swedish manual and interpretation guide, 2nd ed). Vol 2nd ed. Göteborg: Health Care Research Unit, Medical Faculty Göteborg University and Sahlgrenska University Hospital, Göteborg, Sweden; 2002.
62. Sullivan M, Karlsson J, Ware JE. The Swedish SF-36 health survey. 1. Evaluation of data quality, scaling assumptions, reliability and construct-validity, across general populations in Sweden. Soc Sci Med.1995;41(10):1349-58.
63. Alsen P, Brink E, Persson LO. Patients' illness perception four months after a myocardial infarction. J Clin Nurs. 2008;17(5A):25-33.
64. Wilde Larsson B, Larsson G, Larsson M, Starrin B. KUPP- Kvalitet ur patientens perspektiv. 4th ed. Stockholm: Vårdförbundet; 2001.
65. Patton MQ. Qualitative research & evaluation methods. 3rd ed. London: Sage; 2002.
66. Graneheim UH, Lundman B. Qualitative content analysis in nursing research: concepts, procedures and measures to achieve trustworthiness. Nurse Educ Today. 2004;24:105-12.
67. World medical association declaration of Helsinki. Ethical principles for medical research involving human subjects [Internet]. Bulletin of the World Health Organization. 2001;79(4):373-9. Available from: www.who.int/bulletin/archieves/79(4)373.pdf
68. American nurses association. Codes of ethics for nurses with interpretive statements. [Internet] 2011 [cited 2011 april 18] Available from: http://www.nursingworld.org/MainMenuCategories/EthicsStandards/CodeofEthicsforNurses/Code-of-Ethics.aspx
69. Olsen PD, Mahrenholz D. IRB-Identified ethical issues in nursing research. J Prof Nurs. 2000;16(3):140-8.
70. Han SW, McColl E, Barton JR, James P, Steen IN, Welfare MR. Predictors of quality of life in ulcerative colitis. The importance of symptoms and illness representation. Inflamm Bowel Dis. 2005;11(1):24-34.
71. Agreus L, Svardsudd K, Nyren O, Tibblin G. The epidemiology of abdominal symptoms: prevalence and demographic characteristics in a Swedish adult

population. A report from the Abdominal Symptom Study. Scand J Gastroenterol. 1994;29(2):102-9.

72. vanWijk CM, Kolk AM. Sex differences in physical symptoms: the contribution of symptom perception theory. Soc Sci Med.1997;45(2):231-246.
73. Ross S, Walker A, MacLeod MJ. Patient compliance in hypertension: role of illness perceptions and treatment beliefs. J Hum Hypertens. 2004;18(9):607-13.
74. Kuhl EA, Fauerbach JA, Bush DE, Ziegelstein RC. Relation of anxiety and adherence to risk-reducing recommendations following myocardial infarction. Am J Cardiol. 2009;103:1629-34.
75. Leenerts MH, Teel CS. Relational conversation as method for creating partnerships: pilot study. J Adv Nurs. 2006;5:467-76.
76. Leenerts MH, Teel CS, Pendleton MK. Building a model of self-care for health promotion in aging. J Nurs Scholarsh. 2002;34(4):355-61.
77. Larsson IE, Sahlsten MJ, Sjostrom B, Lindencrona CS, Plos KA. Patient participation in nursing care from a patient perspective: a Grounded Theory study. Scand J Caring Sci. 2007;21(3):313-20.
78. Orlando M, Meredith LS. Understanding the causal relationship between patient-reported interpersonal and technical quality of care for depression. Med Care. 2002;40(8):696-704.
79. Wagner EH, Austin BT, Davis C, Hindmarsh M, Schaefer J, Bonomi A. Improving chronic illness care: Translating evidence into action. Health Aff. 2001;20(6):64-78.
80. Polit DF, Beck CT. Nursing research. Principles and methods. 7th ed. London: Lippincott Williams & Wilkins; 2004.
81. Wibeck V. Fokusgrupper. Om fokuserade gruppintervjuer som undersökningsmetod. Lund: Studentlitteratur;2000.
82. Nyström M, Dahlberg K. Pre-understanding and openness – a relationship without hope? Scand J Caring Sci. 2001;15:339-46.
83. Rees G, Fry A, Cull A, Sutton S. Illness perceptions and distress in women at increased risk of breast cancer. Psychol Health. 2004;19(6):749-65.
84. Berglund E, Olsson A. Validering av den Svenska översättningen av The Revised Illness Perception Questionnaire – genom kognitiv intervjuteknik ”think aloud”. Examensarbete (HT07): Sophiahemmet Högskola; 2010.

85. Hagell P, Reimer J, Nyberg P. Whose quality of life? Ethical implications in patient-reported health outcome measurement. Value in Health. 2009;12(4):613-7.
86. Törnvall E, Wilhelmsson S. Quality of nursing care from the perspective of patients with leg ulcers. J Wound Care. 2010;19(9):388-95.
87. Nathorst-Böös J, Munck IM, Eckerlund I, Ekfeldt-Sandberg C. An evaluation of the QSP and the QPP: two methods for measuring patient satisfaction. Int J Qual Health Care. 2001;13(3):257-64.